CRANIOVERTEBRAL JUNCTION REALIGNMENT FOR BASILAR INVAGINATION

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DEPARTMENT OF NEUROLOGICAL SCIENCES

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VELLORE

CERTIFICATE

This is to certify that the dissertation titled "Craniovertebral junction realignment for basilar invagination" is the bonafide original work of Dr. Manikandan, S. N submitted in partial fulfillment of the rules and regulations, for Branch-II M.Ch. Neurosurgery, final examination of the Tamil Nadu Dr. M.G.R. Medical University to be held in August 2010.

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AIM:

- To assess the clinical, functional and radiological outcome after craniovertebral realignment surgery for basilar invagination.

INTRODUCTION:

Basilar invagination is a congenital condition where the tip of the odontoid process invaginates into the foramen magnum. It may result in lower brain stem or upper spinal cord compression producing progressive neurological deficits.

Primary or true congenital basilar invagination may be associated with other vertebral anomalies including occipito-atlantal fusion, hypoplasia of atlas, and hemirings of C1 with spreading of lateral masses, odontoid abnormalities and Klippel-Feil deformity (1).

Acquired basilar invagination or basilar impression is caused by softening of the bone at the base of skull as a result of degenerative disorders such as Paget’s disease of bone, Osteogenesis imperfecta, Hurler’s syndrome, and severe Rheumatoid arthritis or osteoarthritis, tumors or infection.

REVIEW OF LITERATURE:

Development of Craniovertebral junction:

The craniovertebral junction develops from four occipital sclerotomes and from the first and second cervical sclerotomes (1). The occipital bone, clivus and occipital condyles are derived from four occipital sclerotomes. The fourth occipital sclerotome forms the occipital condyles, paracondylar process and the bones surrounding the foramen magnum. The anterior arch of the atlas is derived from a band of tissue, the hypochondral bow (1), which is also derived from the fourth occipital sclerotome. The posterior arch of atlas is derived from both the fourth occipital sclerotome and from the first cervical sclerotome. The atlas ossifies from a single ventral and paired dorsal ossification centres.

The axis is derived from the fourth occipital and the first two cervical sclerotomes. The tip of the odontoid is derived from the fourth occipital sclerotome; the odontoid process from the first cervical sclerotome and body of the axis and dorsal vertebral arch are derived from the second cervical sclerotome. Fusion of the odontoid and axis body begins at 4 years and is completed at the age of 8. Fusion of the apex of the odontoid to the odontoid proper occurs at 12 years.

Developmentally primary basilar invagination may be due to an insufficient amount of paraxial mesoderm, leading to the underdevelopment of the occipital somites causing shortening of the clivus and an enlargement of the foramen magnum in the anteroposterior dimension. During chondrification the cartilaginous dens may transiently reach into the foramen magnum, but descends below the foramen magnum in the fetal period. If this is incomplete, basilar invagination may result (1). Os odontoideum is the dissociation between the odontoid process and the body of the axis whereas the os terminale persistens is the failure of the fusion of the odontoid tip with the remainder of dens (1).

Surgical anatomy of craniovertebral junction

Foramen Magnum:

The foramen magnum has three parts: (a) the squamosal portion which is located in the dorsal aspect of foramen magnum, (b) the basal or clival portion located anterior to the foramen magnum, and (c) the condylar part that connects the squamosal and the clival parts that articulates the atlas lateral mass (2). The hypoglossal canal perforates the skull base at the lateral part of the condyles and transmits the hypoglossal nerve along with the branch of the posterior meningeal artery. The most posterior margin of the foramen magnum is called the opisthion. The anterior most midline of the foramen magnum is termed the basion.

Atlas:

The atlas has two thick lateral masses, which are situated at the anterolateral part of the ring. These are connected in front by a short anterior arch and behind by the long posterior arch. The position of the usual vertebral body is occupied by the odontoid process of the axis. At the base of the posterior arch between the superior facet and the neural arch is a groove for the vertebral artery (2). The first spinal nerve runs parallel to the vertebral artery in this groove. The inferior surface of each lateral mass of the atlas has a circular flat of slightly convex facet which faces downward and medially and articulates with the superior articular facet of the axis. The transverse foramen transmits the vertebral artery upon which the nerve root lies and is situated between the lateral mass and the transverse process. The posterior aspect of the lateral mass of the atlas has become important from a point of view of screw fixation through this strong bone component.

Axis:

The axis is the second cervical vertebra. The odontoid process projects cephalad from its articulation with the axis body. On the ventral odontoid surface is an oval facet, which articulates with the dorsal surface of the anterior atlas arch. In the dorsal aspect of the dens is a transverse groove over which passes the transverse ligament of the atlas. The axis spinous process is large, deeply concave on its caudal border and is the first bifid vertebra in the cervical spine.

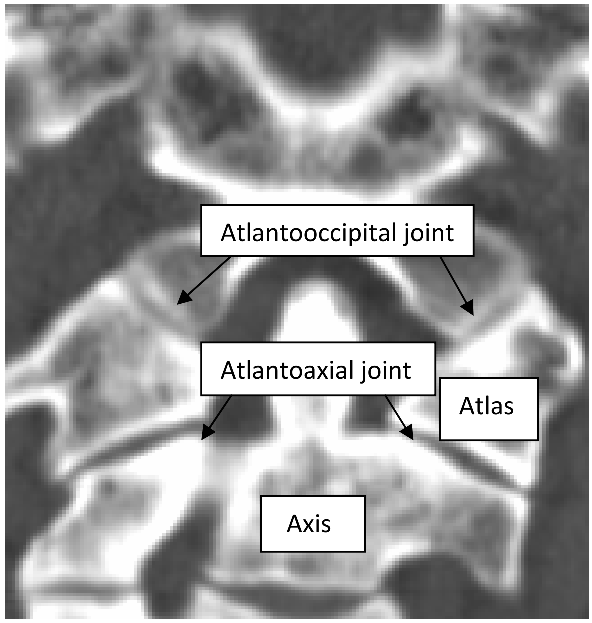
Ligamentous anatomy:

The arrangements of the occipitoatlantoaxial ligaments are to allow for complex motion and to provide stability. The atlanto-occipital joints are prominent, lax and provide poor stability to the joint. The articular capsules of the lateral atlantoaxial facets surround the articular surfaces and are strengthened by atlantoaxial ligaments. The capsules are reinforced by lateral fibers that pass in a rostral direction from the tectorial membrane. A second cervical nerve exits from the vertebral canal immediately dorsal and adjacent to the joint capsule. Two central atlantoaxial joints are located between the dens and the transverse ligament dorsally and between the dens and the anterior atlas arch ventrally. There are ligaments between the anterior arch of the atlas and the dens and behind it. The cruciate ligament has a vertical component that attaches to the rim of foramen magnum in the midline and inferiorly to the midportion of the body of the axis. The apical ligament of the dens extends from the rim of foramen magnum to the dens. The alar ligament is a separate portion that swings from the lateral anterior rim of foramen magnum and comes toward the dorsal aspect of the dens. The transverse portion of the cruciate ligament is approximately 6–7 mm in height and is one of the most important ligaments of the body. It is attached on either side to a tubercle in the inner ring of the atlas lateral mass and crosses from side to side in a dorsal convex arch to divide the atlas ring into a dorsal and ventral component. The ventral component contains the odontoid process and the dorsal component encompasses the spinal cord and the spinal accessory nerves. The ligament presents a fibrocartilaginous surface allowing for free gliding motion to occur over the posterior facet of the dens. The tectorial membrane is dorsal to the cruciate ligament and a strong band of longitudinally oriented fibers that are attached to the dorsal surface of C3 vertebrae, the axis body and to the body of the dens. The ligament ascends upward and widens to attach to the base of the occipital bone. This membrane is the rostral extension of the posterior longitudinal ligament of the vertebral column. Ruptures of the cruciate ligament are easily identified and can aid in the decision making of craniocervical stability.

Biomechanics of craniovertebral junction:

The occipitoatlantoaxial complex (Figure 1) is the most mobile of the axial skeleton. This functions as a single unit with the axis being interposed between the skull and cervical spine. Flexion and extension movements occur at the occipitoatlantal and the atlantoaxial articulations. This accounts for 25% of the flexion and extension movement in the neck. In children, the anteroposterior translation that occurs between the anterior arch of the atlas and the dens or the so-called predental space can be up to 5 mm until the age of 8 years, and in adults, the predental space should be less than 3 mm (3). With disruption of the cruciate ligament, the load is then placed on the alar and apical ligaments, which quickly become incompetent.

Figure 1 Coronal view of a CT scan showing Occipitoatlantoaxial complex



The largest degree of rotation occurs at the atlantoaxial joint (Figure 1). Usually, a rotation past 25–30° brings the middle and lower cervical segments into play. This prevents increased rotation at the atlantoaxial joint. Anatomic studies have shown that stretching and kinking of the contralateral vertebral artery occurs between 30 and 35° of atlantoaxial rotation (2). When rotation exceeds 40°, an interlocking of the facets occurs between the atlas and the axis vertebra. When an acute rotation of the atlantoaxial joint is made exceeding 45°, the ipsilateral vertebral artery may demonstrate angulation and occlusion. This has particular significance in children with atlas assimilation, with individuals participating in football and wrestling, and those who undergo excessive rotation of the head during general anesthesia or forceful head manipulations (2). The unique anatomic configuration of the craniovertebral junction creates a distinct biomechanical behavior that differs from that of other spinal joints. There is no intervertebral disc between the occiput and C1 and C2. The ball-and-socket shaped occiput-C1 joint allows slightly more flexion-extension than the other levels of the spine. The biconvex articular surfaces of the C1-2 joints allow gliding and wide rotation of the C1 around the dens. The atlanto-axial joint is more flexible and allows more than 90 degrees of rotation bilaterally. When the transverse component of the cruciate ligament has been disrupted, the alar ligaments are still intact. Hence the amount of displacements remains between 5 and 6 mm until the alar ligaments become incompetent (2). The transverse atlantal ligament is the strongest and thickest ligaments of the entire spine.

Pathogenesis of basilar invagination:

Several theories have been suggested to clarify the probable cause and origin of basilar invagination. They include mechanical, embryological dysgenesis, genetic abnormalities or viral infections (4). Goel et al speculated that basilar invagination is secondary to an abnormally inclined alignment of the facets of the atlas and axis (4). The progressive slippage of the atlas over the axis secondary to this malalignment, results in invagination of the odontoid process into the foramen magnum (5).

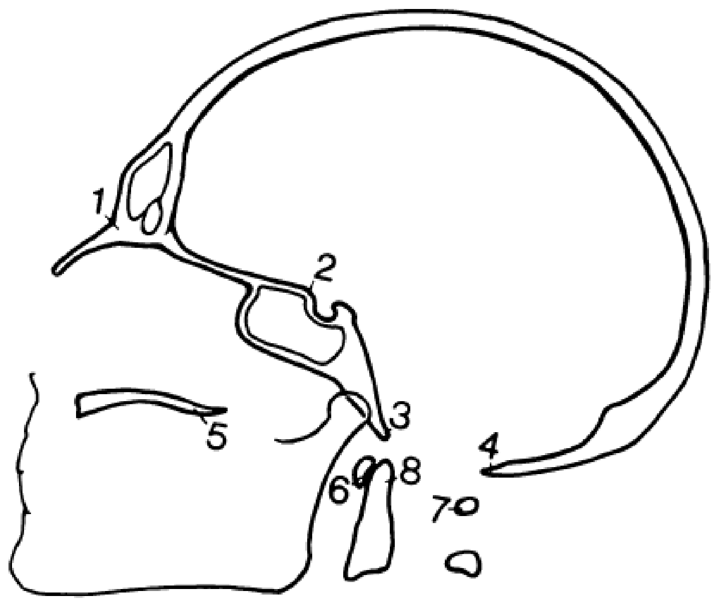
Classification of basilar invagination:

Goel et al (4) classified basilar invagination into Group I and Group II. Group I included invagination of the odontoid process into the foramen magnum with compression of the brainstem. The angle of the clivus and the posterior cranial fossa volume are unaffected in these patients. In Group II, on the other hand, the assembly of the odontoid process, anterior arch of the atlas and the clivus migrates superiorly resulting in reduction of the posterior cranial fossa volume, which is the primary pathology in these patients. The Chiari malformation or herniation of the cerebellar tonsil is considered to be a result of reduction in the posterior cranial fossa volume. The same author later proposed another classification to approach these patients surgically, Group A and Group B (4). In Group A basilar invagination there is a 'fixed' atlantoaxial dislocation and the tip of the odontoid process invaginates into the foramen magnum and is above the Chamberlain line, McRae line and Wackenheim's clival canal line and resulted in direct compression of the brainstem. In Group A some patients have Chiari malformation, and this feature differentiates the newer classification from the earlier classification where there was no Chiari malformation. In Group A, the atlantoaxial joints are 'active' and their orientation was oblique as compared to horizontal orientation normally. In Group B basilar invagination the odontoid process and clivus remained anatomically aligned despite the presence of basilar invagination and other associated anomalies. In Group B, the tip of the odontoid process is above Chamberlain's line but below McRae's and Wackenheim's line, the atlantoaxial joints are normal and are normally aligned.

Radiological criteria:

The bony landmarks of the skull base and the craniovertebral junction are shown in Figure 2

Figure 2



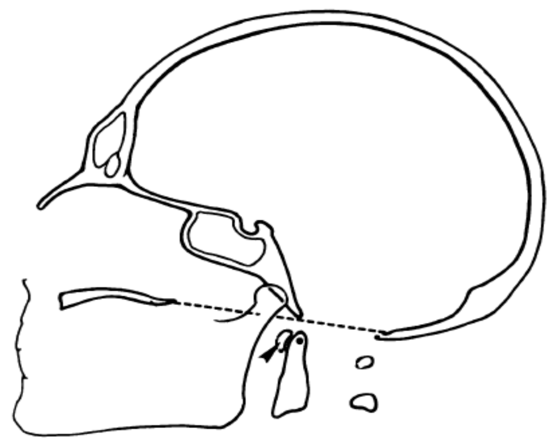
1. Nasion
2. Tuberculum sella
3. Basion
4. Opisthion
5. Posterior end of hard palate
6. Anterior arch of atlas
7. Posterior arch of atlas
8. Odontoid.

Chamberlain's line:

Chamberlain’s line (Fig 3) extends from the posterior pole of the hard palate to the opisthion (6). Mc gregor’s line extends from the posterior pole of the hard palate to the lowest point of the occipital squamousal surface. McRae’s line is between basion and opisthion (7).

Figure 3

Chamberlain’s line



Basilar invagination is diagnosed when the tip of the odontoid process was at least 2 mm above Chamberlain's line (4).

Mc gregor’s line:

Mc gregor’s line extends from the posterior end of hard palate and lowest point of the occipital squamousal surface. The basilar invagination is diagnosed when the tip of the odontoid is 1 mm ± 3.6 to 6.6 mm above this line (8).

McRae line:

McRae line is between basion and opisthion. The odontoid tip lies below this line in normal subjects and if the odontoid tip is above this line then it is diagnostic of basilar invagination.

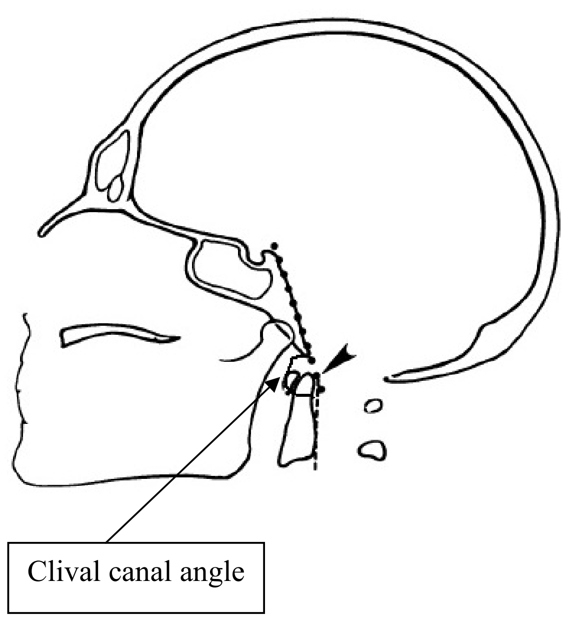
Atlanto-dental distance:

The distance between anterior arch of atlas and the odontoid is the atlanto-dental distance. The atlanto-dental or atlanto-axial distance more than 5 mm is considered as abnormal in children and more than 3 mm is considered as abnormal in adults.

Wackenheim's clival line:

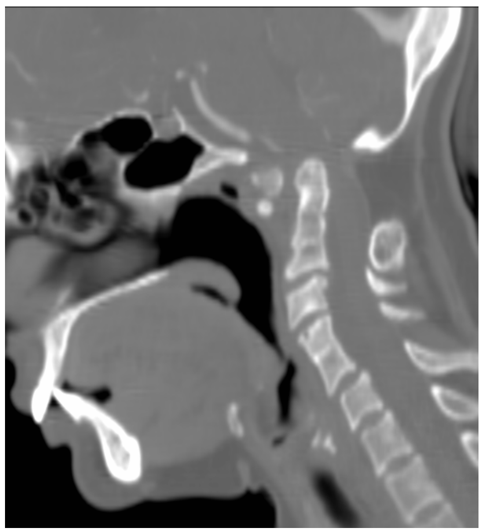
Wackenheim clivus baseline (Fig 4) is a line along the clivus extrapolated inferiorly into the upper cervical spinal canal (9). The angle formed at the intersection of the Wackenheim clivus baseline with a line constructed along the posterior surface of the axis body and odontoid process is the clivus canal angle. It normally ranges from 150 in flexion to 180 degrees in extension.

Figure 4: Wackenheim clival baseline and clival canal angle



The tip of the odontoid process is significantly superior to Wackenheim's clival line in Group A patients. In Group B patients, the relationship of the tip of the odontoid process and the lower end of the clivus and the atlanto-dental interval is normal. In Group B patients (4) the basilar invagination is due to the rostral positioning of the plane of the foramen magnum in relation to the brainstem as shown in the Fig 5.

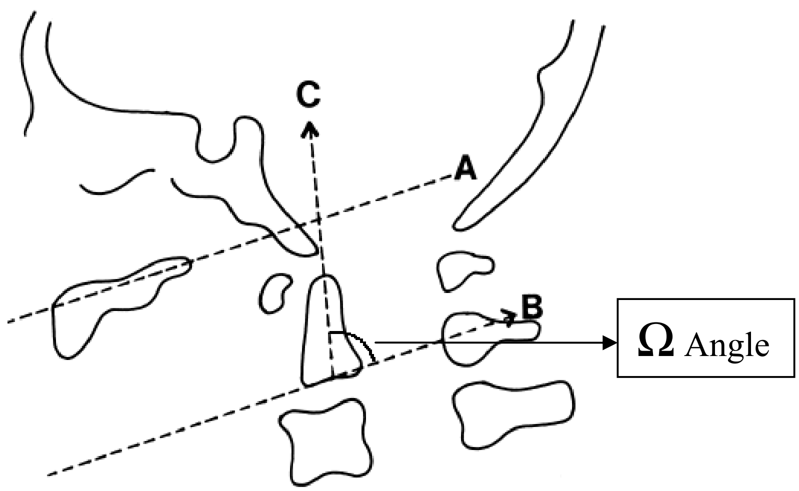
Figure 5: Imaging showing Group B type of basilar invagination



Omega angle:

Goel et al described a modified Omega angle as a measurement of the angle of the odontoid in the sagittal plane in relation to the plane of the hard palate (10). A vertical line is drawn traversing through the centre of the base of the axis parallel to the line of the hard palate. The line of the hard palate is unaffected by the relative movement of the head and the cervical spine during the movement of the neck in these 'fixed' craniovertebral anomalies (10). The Omega angle (Fig 6) is the angle between the line B and line C. The Omega angle is severely reduced in Group A patients while it is much larger in Group B patients.

Figure 6 : Modified omega angle



METHODS OF TREATMENT OF BASILAR INVAGINATION

Transoral odontoidectomy:

The transoral approach for craniospinal malformations was first used by Kanavell and Le Fort in 1918, Fang and Ong in 1962, Greenberg et al in 1968, Grison in 1967, Fokes and Jomin and Bouasakao in 1977, Menezes et al in 1980, and more recently by Crockard in 1985 (11).

Procedure

A Spetzler-Sonntag retractor is placed by retracting the tongue and endotracheal tube to allow maximum exposure of the posterior pharyngeal area. The uvula and the soft palate are retracted superiorly with a red rubber catheter inserted through the nose. This retraction improves exposure of the upper portion of the posterior pharyngeal wall overlying the tip of the odontoid and prevents secretions from running into the incision. The posterior pharynx is then infiltrated with 1% lidocaine with epinephrine. The incision is typically 1.5 to 2 cm in length and is carried through the posterosuperior pharyngeal constrictor muscle in the midline raphe with "no touch" oral cavity technique.

A Crockard self-retaining retractor is then placed in the midline pharyngeal incision and spread laterally to expose the anterior arch of C1. The fluoroscope is used to confirm anatomic landmarks. Once the arch of C1 has been exposed anterior arch of C1 is drilled to expose the anterior portion of the odontoid process followed by detachment of the apical and alar ligaments at the top of the odontoid process. Then the C2 body and the odontoid are drilled to relieve the compression. Transoral surgery causes instability of C 1-2 in 70% of patients which requires fusion (12). After decompression, occipitocervical fusion is being done either on the same day or after 7 days of traction (13). The total angular range of motion increased significantly during flexion, extension and lateral bending but not during axial rotation. This was studied in vitro by Dickman et al (14) who showed spinal stability is mandatory to prevent the acute or delayed effects of transoral odontoidectomy (14). Cervical fusion alone may not be possible when the ring of C1 is fractured, C1 incorporated into the occiput or in patients with rheumatoid arthritis; in this case occipitocervical fusion is the preferred method of stabilization.

Results of transoral odontoidectomy:

Jain et al (15) studied the surgical outcome of 74 patients, who underwent transoral decompression for ventral irreducible craniovertebral junction anomalies to evaluate the perioperative complications and problems encountered. The patients had irreducible atlantoaxial dislocation (n=24), basilar invagination (n=16), and a combination of both (n=35). Following surgery, occipitocervical stabilization was carried out in 50 (67.5%) and atlantoaxial fusion using Brooks' construct in 18 (24.3%) patients. The major morbidity included pharyngeal wound sepsis leading to dehiscence (20.3%) and hemorrhage (4%), velopharyngeal insufficiency (8.1%), CSF leak (6.7%) and inadequate decompression (6.7%). Neurological deterioration occurred transiently in 17 (22.9%) and was sustained in 7 (9.4%) patients. The mortality in six cases was due to operative trauma, exsanguinations from pharyngeal wound (one each), postoperative instability and inability to be weaned off from the ventilator (two each). Of the 47 (63.5%) patients at follow up ranging from 3 months to 2 years, 26 (55.3%) showed improvement from their preoperative status while 14 (29.8%) demonstrated stabilization of their neurological deficits. Seven (14.9%) of them deteriorated. Jain et al concluded transoral odontoidectomy has logical and effective in relieving ventral compression due to craniovertebral junction anomalies, but they have also added that it carries the formidable risks of instability, incomplete decompression, neurological deterioration, CSF leak, infection and palatopharyngeal dysfunction.

Menezes et al (16) studied 72 patients between the ages of 6 and 82 years who underwent ventral transoral transpharyngeal decompression of the craniocervical junction. Pluridirectional lateral tomography of the CVJ was obtained 7 days after surgery to determine craniovertebral stability. This was done in the flexed and extended positions, as well as with and without traction. Of the 72 individuals who underwent a ventral decompression, 52 patients showed instability and required a dorsal fixation procedure. All patients showed neurological improvement. Six individuals who were ventilator-dependent following either trauma or a previous primary posterior decompression had resolution of their neurological symptoms and signs in the postoperative period. Downbeat nystagmus, sleep apnea and brain-stem signs were prominent features in 15 individuals with basilar invagination and the Chiari malformation. These signs regressed following the ventral decompressive procedure. Two patients died within the 1 st month of operation, one due to a myocardial infarction and other due to Escherichia coli septicemia from a urinary tract infection. One patient had a postoperative pharyngeal wound infection and a retropharyngeal abscess that required drainage.

Goel et al (10) performed transoral surgery in 99 patients (Group I, 78 patients [89%] and in Group II, 21 patients [21%]. Group I included invagination of the odontoid process into the foramen magnum with compression of the brainstem. In Group II, the assembly of the odontoid process, anterior arch of the atlas and the clivus migrates superiorly resulting in reduction of the posterior cranial fossa volume. Following transoral surgery, in six cases homologous bone graft was placed between the residual C2 body and the inferior part of the clivus to assist fusion. In three patients a transoral plate and screw fixation of the clivus to the cervical vertebral body was performed. The bone graft was placed underneath the metal plate. These patients were placed in halo fixation following the surgery. This anterior fixation was abandoned due to infection, rejection of the metal implant and poor visualization. Dorsal fixation was performed in the same surgical session following a transoral surgical procedure in 18 Group I patients. In these cases the indication for immediate fixation was relatively high mobility of the cervical vertebral bodies during drilling. In 39 other Group I patients, fixation was performed as a second stage surgery. Excessive pain and spasm of the neck muscles and suboccipital radicular pain formed the primary indication for fixation in these patients. No patient worsened in motor function prior to second-stage fixation. In this group fixation was performed after the initial surgery within 15 days in 16 patients, within 2 months in 11 patients, and between 2 and 6 months in 12 patients. In Group II, a posterior fixation procedure was conducted following transoral decompression in the same surgical sitting in one patient. In four patients fixation was performed within 2 weeks after transoral surgery. No patient needed a fixation procedure as a delayed measure. In six Group II patients, no fixation was necessary, even after both anterior and posterior decompressive operations. They concluded that the transoral surgery is indicated in Group I patients whereas Group II patients warrants foramen magnum decompression only.

Disadvantages of transoral odontoidectomy:

Drawbacks of the approach include a limited operative view, a deep working distance, contamination by normal oral flora (17, 18) of the oral cavity, dehiscence of the posterior pharynx, alteration in phonation secondary to effects of surgery on the pharynx (19, 20), tongue edema (21), the potential need for prolonged intubation (21), and the requirement of avoiding oral intake to allow the pharyngeal closure to heal (20). The major morbidities include vertical occipitoatlantal subluxation with vertebral artery occlusion and brain stem stroke (22). CSF leaks encountered during the course of a transoral surgery have potentially devastating consequences. Meningitis caused by oral bacteria invading the CSF and death have been reported with this technique (18).

The worsening of basilar invagination as a cause of failure of transoral odontoidectomy has been reported earlier (12, 23). Transoral odontoidectomy causes further cranial settling of the upper cervical spine (C2 body) causing brain stem compression. This is caused by the horizontal separation of the lateral mass of C1 due to removal of anterior arch and ligaments. A partial resection of anterior arch of C1 will minimize horizontal separation of lateral mass and thus cranial settling of C2. Such a worsening has been seen even when a posterior fixation was performed with wires as the pullout strength of wire is less than the screw (23).

Neurological deterioration after transoral odontoidectomy:

Fifty patients in the series reported by Jain et al (15) were preoperatively in Nuricks grade III or IV, being partially or totally dependent on others for their daily needs. Of the 47 (63.5%) patients at follow up ranging from 3 months to 2 years, 26 (55.3%) showed improvement from their preoperative status while 14 (29.8%) demonstrated stabilization of their neurological deficits. Seven (14.9%) of them deteriorated. Thus, 21 of the 47 (44.6%) patients seen at follow up did not show significant neurological recovery. Twenty four patients had a significant respiratory compromise. The repetitive trauma due to craniovertebral anomalies leads to anterior horn cell destruction, gliosis of gracile and cuneate nuclei and demyelination of the corticospinal tracts and posterior columns. Stagnant hypoxia secondary to venous stasis or occlusion of the vertebral or spinal arteries and preexisting microscopic intracranial abnormalities also contribute to the neural damage (15). However, even minor trauma on an already compromised cord may cause respiratory deterioration. The common features in patients who deteriorated and those who could not be weaned off from the ventilator following transoral surgery were the presence of advanced spastic quadriparesis and respiratory compromise. On MRI, they showed evidence of marked spinal cord compression with thinning of the cord and hyperintense cord signals.

Tuite et al (24) found that the higher rate of neurological morbidity may be related to greater severity of preoperative neurological deficits. One of patients reported by Jain et al (15) had transient haemodynamic instability and bradycardia during transoral odontoidectomy. He developed quadriplegia with respiratory arrest following reversal from anaesthesia. This clinical syndrome of complete cervicomedullary transection could have been due to accentuation of cord damage by the recurrent posterior displacement of the odontoid while drilling (15). However, intramedullary hemorrhage from the sulcal branches of anterior spinal artery due to the sudden release of pressure cannot be ruled out in these patients (15). Spinal cord injury during the sublaminar passage of wires or instability of CVJ during repositioning for posterior stabilization may also add to neurological injury (24). To prevent the cord injury while drilling Jain et al (15) implicated a lateral rather than a downward pressure should be applied and a thin posterior cortical surface of odontoid should be left which can be removed after elevating it from the posterior longitudinal ligament. Neurophysiologic monitoring with evoked somatosensory or brainstem auditory potential helps in predicting potential brain-stem or cord injury (15).

Cranial Settling:

The unique anatomy and biomechanics of the CVJ differentiate this region from other spinal segments. Naderi et al (23) reported further cranial settling in two patients whom underwent transoral decompression and occipitocervical fusion which necessitated a second decompressive procedure in one of the cases. The other patient was asymptomatic, and an osseous fusion was demonstrated between the C-2 vertebral body and lower aspect of the clivus. Transoral odontoidectomy results in a severe ligamentous and osseous destruction and it alters the CVJ anatomy and affects the biomechanics of the region. Both these patients underwent C1 anterior arch excision which probably caused further cranial settling in these patients. The author in another study (25) demonstrated the effects of odontoidectomy in a cadaveric model by compressing the occiput–C3 complex before and after resection of the anterior arch of C-1. In the specimen in which the C-1 anterior arch had been sectioned, horizontal separation of the lateral masses of C-1 occurred and resulted in further cranial settling of the C-2 body. The author determined that the preservation of C-1 anterior arch and lamina minimizes the horizontal separation of the C-1 lateral masses.

The other aspect involved in CVJ instability is the choice of fixation technique. The most advantageous biomechanical results were demonstrated when using C1–2 transarticular screws or C-2 pedicle screws in association with occipital fixation (24) instead of wires. Endoscopically assisted transoral surgery represents an emerging alternative to standard microsurgical techniques for transoral approaches to the anterior cervicomedullary junction. Frempong Boadu et al (26) described a series of seven consecutive patients treated with endoscopically assisted transoral surgery for decompression of high cervical and clival abnormalities. Successful decompression was achieved in all seven patients. There were no adverse neurological sequelae. One patient died from a perioperative myocardial infarction. At a mean clinical follow-up of 6.16 months, neurological status was noted to be stable or improved in all remaining patients. Some of the transoral series are summarized in table no 1.

Author	No of patients	Preoperative Traction	Improved/ Stabilized neurological status	Complications & Incidence	Hospital stay	Follow up details
Menezes (16) et al 1998	72	No	All	2 mortality due to MI and Septicemia	N.A	N.A
Mark (22) et al 1989	53	No	All	Morbidity-3 (6)% -wound dehiscence, brainstem stroke, CSF leak & mortality-3 (6%)-pneumonia and pulmonary embolus.	N.A	24 months (median)
Laborde (11) et al 1992	15	No	Not available	Morbidity-12 (80%), atlantoaxial dislocation, CSF leak, hemorrhage, infection & hydrocephalus Mortality-2 (13.3%), infection & hemorrhage	N.A	N.A
Goel (10) et al 1998	99	Yes	All	Morbidity- 1(1%)	N.A	2 months to 9 years (average 43 months).
Jain (15) et al 1999	74	Yes	55.3% improved 29.8% stabilized	22.9% - deterioration in neurological status Dehiscence (20.3%) and hemorrhage (4%), Velopharyngeal insufficiency (8.1%), CSF leak (6.7%) and inadequate decompression (6.7%). Mortality-6.1%	N.A	47 (63.5%) patients were followed up for 3 months to 2 years
Landeiro (27) et al 2007	38	No	36.8% improved 60.2% stabilized	Morbidity-18.4% due to dehiscence, pulmonary infection & CSF leak. Mortality-1(2.6%)	N.A	N.A

Table 1 Major Transoral series and their results:

Transcondylar approach for resection of the dens:

Al-Mefty et al (28) introduced the transcondylar approach for resection of the dens as an alternative to the transoral approaches. Ture et al (28) described modifications of transcondylar approach for resection of dens via transatlas route advantage being preserving the jugular bulb, hypoglossal nerve, and facial nerve and able to perform the occipital condyle–C2 fusion is one stage. This extreme lateral–transatlas approach was used for the resection of the dens of the axis in five patients. Unilateral occipitocervical fusion was performed during the same procedure. There were no cases of intra- or postoperative complications. There were no cases of postoperative infections, wound infection, or CSF leakage. The follow-up period ranged from 13 to 24 months (median 17.2 months) within which no craniocervical instability was demonstrated.

Halo traction and fusion:

Simsek et al (29) report CT and MRI compatible halo traction for a patient with basilar invagination was treated with traction followed by posterior fusion. The patient underwent halo traction and at the end of 4 weeks she underwent a posterior fusion. She was on halovest traction for a period of 3 months postoperatively and was removed at the end of 4 months when her JOA score improved by six. She had an acute neurological deterioration one year later after a trauma. She was again treated with the same halovest traction technique and her occipital screws were tightened. Her neurological status improved on the first postoperative day. At one year follow-up after the second surgery there was neither neurological deterioration nor implant failure. Menezes (29) described significant improvement in basilar invagination and atlantoaxial subluxation cases with traction treatment. Joseph and Rajshekhar (30) reported a patient who presented with basilar invagination, Chiari formation, and syringomyelia, in which the anomalies resolved under cervical halter traction therapy for 4 weeks without any operative intervention. Kyoshima et al (29) was the first to report a simple cervical traction method with the halo vest apparatus for the unstable CVJ injuries. Moreover, bed rest is not necessary during the procedure; it may be an advantageous point for preventing deep venous thrombosis and pulmonary embolism, particularly in elder patients. However Goel et al (10) performed a posterior fixation procedure in a reduced position of the basilar invagination and the atlantoaxial dislocation following cervical traction in four patients. All these patients needed a transoral surgery at a later stage because the reduced position could not be maintained by the implant.

Craniovertebral realignment for basilar invagination:

Goel et al (31) proposed that basilar invagination is a result of abnormally inclined facets of the atlas and axis caused by congenital malformation of the bones in the region. Progressive worsening of basilar invagination and atlantoaxial dislocation is probably secondary to increasing "slippage" of the atlas over the axis (31). This slippage can be severe enough to cause spondyloptosis of the atlas over axis. The joint in these cases is not "fixed" or "fused" but is mobile and, in some cases, is hypermobile and is probably the prime cause for basilar invagination (5). They thus proposed a distraction surgery in which the C2 is forcibly brought down through skull traction and was kept in place with a plate and screw. They reported 3 cases where there was progressive neurological detoriation after transoral surgery which improved after distraction and fusion of C1-2 joints (5). In this surgery by atlantoaxial joints are opened bilaterally, the articular cartilages drilled and filled with a spacer connected to the vertical plates that are screwed on to the C1 lateral mass.

Surgical procedure described by Goel et al (31)

The patients are placed in cervical traction prior to induction of anesthesia, and the weights are progressively increased to approximately one fifth of the total body weight. The patient is positioned prone with the head end elevation to 35°. The atlantoaxial facet joints are approached via the pars of C2 and exposed after sectioning of the large C-2 ganglion. The joint capsule is excised and the articular cartilages are removed with a micro drill. The joint is distracted bilaterally. The pieces of corticocancellous bone with metal plate spacer are used as strut graft and are packed into the joints. The size of the spacers depends on the space available within the distracted joint space. Posteriorly bone graft was placed between the posterior elements of C1–suboccipital bone complex and C-2 after decorticating the host bone. The neck is immobilized in a Philadelphia collar for 3 months. The patients were followed up with MRI imaging 7 days postoperatively and after a follow up of every 6 months.

Goel et al compared basic craniovertebral craniometry pre and postoperatively (31). He found that the odontoid process, the clivus as well as the entire CVJ alignment were improved after surgery. The tip of the odontoid process descended in relation to the Wackenheim clival line, Chamberlain, and MacRae lines, indicating reduction in basilar invagination. The posterior tilt of the odontoid process, as indicated by the modified omega angle, decreased postoperatively. Reduction of the basilar invagination and atlantoaxial dislocation was achieved in all patients. The follow-up period ranged from 1 to 4 years (mean 28 months). Symptoms improved to varying degrees in all cases following surgery, and all the patients were independent. There were no intra- or postoperative vascular, neurological, or infection complications. No patient suffered a delayed neurological worsening sufficient to warrant a transoral or a posterior decompressive surgery or any other type of operative procedure. No patient required a reexploration for failure of implant fixation. Immediate postoperative and follow-up imaging confirmed fixation and fusion as well as reduction of the basilar invagination in all cases. Fusion was considered successful when the implant was shown to maintain the distraction and reduction of the basilar invagination on dynamic radiography 6 months after surgery. Successful and sustained distraction and reduction of basilar invagination was observed in all patients. Torticollis improved significantly following surgery in all patients and in four patients there was a complete symptomatic recovery. On examination there was at least some degree of C-2 sensory loss in all cases.

Distraction and fusion for basilar invagination with syringomyelia:

Goel et al (32) described 12 patients in whom syringomyelia was associated with congenital bony anomalies including basilar invagination and fixed atlantoaxial dislocations. Eight had Chiari malformation. All underwent atlantoaxial manipulation and restoration of the craniovertebral region alignment. No patients underwent a posterior foramen magnum decompression. Following surgery all patients showed improvement and restoration of craniovertebral alignment during follow up period of 20 months (mean 7 months). Distraction and fusion was considered to be the optimal treatment for patients with osseous anomalies associated with syringomyelia even though the radiological improvement of syringomyelia could not be evaluated because of implants.

Distraction and fusion for rheumatoid arthritis:

Goel et al (33) also reported a case series of 9 patients of rheumatoid arthritis with basilar impression treated with the same surgical strategy of distraction and fusion. Follow up range was 4 to 48 months (mean 28 months). None suffered a delayed neurological worsening and none required a reexploration for failure of implant fixation. Immediate postoperative and follow-up radiography confirmed fixation and fusion as well as reduction of the basilar invagination. The authors speculated that the main pathogenesis of basilar invagination is an abnormally inclined position of C1-2 joint as a result of congenital bone abnormality, and progressive worsening of the dislocation is likely secondary to increasing subluxation of C-1 onto C-2.

Posterior fusion Of C1and C2 and their biomechanics:

Gallie fusion and modified Gallie's fusion:

Gallie first described posterior C1-C2 sublaminar wire fixation in 1939 with the use of a steel wire. In the Gallie technique (34), a single autograft harvested from the iliac crest is notched inferiorly and placed over the C2 spinous process and leaned against the posterior arch of C1. The graft is held in place by a sublaminar wire that passes beneath the arch of C1 and then wraps around the spinous process of C2. Passage of the sublaminar wire under the lamina of C2 is avoided in order to decrease the risk of neural or dural injury. The Gallie fusion offers good stability in flexion and extension. However, like interlaminar clamping it offers very poor stabilization during rotational maneuvers. Consequently, the rate of nonunion with the Gallie fusion alone has been reported to be as high as 25% (35).

The Gallie technique was modified by Sonntag in the early 1990s. Sonntag's modified technique (34) improves the rotational stability of the Gallie fusion technique while avoiding the bilateral sublaminar C1-C2 cable passage of the Brooks-Jenkins technique. In the Sonntag technique, (34) a sublaminar cable is passed under the posterior C1 arch from inferior to superior. Next a notched iliac crest is placed in between the spinous process of C2 and wedged underneath the posterior arch of C1 unlike the Gallie technique where the bone graft is notched over the spinous process of C2 and simply leaned onto the posterior arch of C1. Both the superior aspect of the C2 spinous process and the inferior arch of C1 are decorticated before graft placement. The cable is then looped over the iliac crest autograft and placed into a notch created on the inferior aspect of the C2 spinous process. The cable is then tightened and crimped. In patients treated with a wiring procedure only, Sonntag recommends the use of a halo to immobilize patients for three months after surgery and the use of a rigid cervical collar for an additional one to two months after that. With this kind of immobilization he has demonstrated a 97% fusion rate with the technique (34).

Brooks-Jenkins fusion:

In the Brooks-Jenkins fusion technique, unlike the Gallie fusion technique, two separate iliac crest autografts are placed between C1 and C2. Each autologous iliac crest graft is beveled superiorly and inferiorly and wedged in between the C1 and C2 lamina on each side of the midline. One sublaminar cable is then passed on each side of the midline under both the C1 and C2 arches and wrapped around each bone graft respectively. The cables are then tightened around the grafts and secured and crimped in place. The Brooks-Jenkins fusion technique provides more rotational stability than does the Gallie technique (34). The rate of fusion after this technique has been reported to be as high as 93% and is improved by the use of halo immobilization. The disadvantages of the Brooks-Jenkins fusion technique include the need for passage of bilateral sublaminar cables beneath both C1 and C2. This carries a higher potential rate of neurological or dural injury than does the single cable passage under the posterior C1 arch for the Gallie technique.

Atlantoaxial fixation biomechanics:

The major posterior fixation methods include various bone graft and wiring techniques, atlantoaxial screw fixation, and interlaminar clamps. Posterior wiring of the atlas and axis with the incorporation of a bone graft has been described with various modifications by Gallie, Brooks and Jenkins, and Papadopoulos et al. Monofilament wire has been replaced by a variety of more flexible and stronger cable systems. Biomechanical studies examining the stabilizing potential of internal fixation of C1 and C2 are generally compared to posterior wiring and graft techniques.

Hanley and Harvell (36) evaluated the immediate stability of midline, Gallie, and Brooks wiring techniques in a spinal injury model consisting of a type II odontoid fracture and transected transverse ligament. All methods restored the stability of the injured segment to at least the level of the intact specimen when tested in flexion, extension, and rotation. The Brooks fixation, however, resulted in the stiffest stabilization, being at least twice as stiff as the midline wiring procedure or the Gallie technique.

Grob et al (37) compared four different methods of atlantoaxial stabilization: wire fixation with a midline graft (Gallie-type), wire fixation with two laminar grafts (Brooks-type), transarticular screw fixation with a midline bone graft and interspinous process wiring (Magerl technique), and bilateral laminar clamps with a midline graft (Halifax technique). After creation of a soft tissue type injury consisting of transaction of the alar, transverse, and capsular ligaments, ten cadaveric spines were stabilized with these four techniques applied in random fashion. After fixation, motion stability was assessed in flexion/extension, lateral bending, and axial rotation. In the intact specimens, the mean range flexion across this segment was 12.7°. After injury, sagittal plane rotation increased to 30.2°. The Gallie procedure, however, provided significantly less stability in flexion, extension and axial rotation and lateral bending. Magerl C1–C2 transarticular screws provided the greatest stability in axial rotation.

Occipitocervical fixation biomechanics:

Fixation from the occiput to the atlas is more challenging from a biomechanical aspect than fixation of just the C1–C2 segment. Numerous different techniques of achieving internal fixation of the entire CVJ have been biomechanically evaluated. Oda et al (38) assessed five different occipitoatlantoaxial fixation techniques using an odontoidectomy model. The techniques can be roughly divided into three groups: semi-rigid fixation using a loop attached to the skull with wires placed through burr holes and to C1 and C2 with sublaminar wires, rigid fixation of the occiput with screws and semi-rigid fixation of the spine with C2 claw hooks, and rigid fixation using occipital screws combined with rigid spinal fixation using either C1transarticular screws or C2 pedicle screws. While all the techniques significantly increased stiffness in sagittal plane rotation compared with the injured state, the rigid fixation techniques were significantly stiffer than the semi-rigid wiring construct.

Disadvantages of occipitocervical fusion:

Moorthy et al (39) assessed the changes in the cervical spine curvature following occipitocervical fusion in pediatric population by measuring the sagittal curvature and the whole cervical spine alignment in the preoperative, immediate postoperative and follow-up radiographs in 14 patients. At a mean follow-up of 16 months eleven patients (79.5%) demonstrated a hyper lordotic curvature. The mean angle of sagittal curvature in the immediate postoperative period was 22 $\pm$ 10 .1 degrees and this showed a statistically significant increase to 35.9 $\pm$ 18 degrees. Nine patients underwent removal of the implants and wires to reduce the hyperlordosis. Seven of the nine patients were available for long term follow-up (mean 28.3 months). The mean change in the angle at follow-up was 4.6 $\pm$ 3 degrees which was not statistically significant. This was because of "crankshaft phenomenon" related to the restriction of growth posteriorly and uninterrupted growth of the vertebral end plate anteriorly and neurocentral synchondrosis. It is argued that the newer methods of stabilization can prevent the occurrence of the crankshaft phenomenon. But in patients with occipitalized atlas it may be not be possible to perform C1-2 stabilization alone. The authors recommend a prophylactic removal of the implant once bony fusion (between 6-12 months of surgery) is achieved to stabilize the angle of sagittal curvature in these patients.

METHODS AND MATERIALS:

Patients:

This prospective study included all consecutive patients with basilar invagination and impression in one neurosurgery unit of CMC Vellore admitted from April 2007 to February 2009 who had at least 6 months follow-up except 2 patients. Patients with irreducible atlanto-axial dislocation were excluded. Thus 20 patients, 14 males and 6 females were included in this study. 17 patients with basilar invagination were in Group I and three with basilar impressions were in Group II.

Preoperative evaluation:

- *Clinical:*

The various symptoms with the durations were noted. Detailed neurological status of the patients was assessed including ability or inability to walk with or without support, involvement of sensory tracts, autonomic functions and the cranial nerve dysfunction. The Nuricks score and the modified Japanese orthopedic association scores were assessed before surgery. The proforma used is shown as Appendix 1.

- *Plain X-rays:*

  Flexion, extension and neutral X-rays of craniovertebral junction were done.

- *3D CT angiogram:*

  The 3D CT angiogram of the craniovertebral junction was done for all patients with sagittal and coronal reconstructions. The position of the vertebral artery, thickness of the C2 pedicle, the width of the C1 lateral mass were noted in all patients to plan for surgery.

- *MRI:*

MRI of craniovertebral junctions was done in all patients to assess the degree of compression at the upper cervical cord. The narrowest sagittal canal diameter at the level of the upper cervical cord and the presence or absence of Chiari malformation was noted.

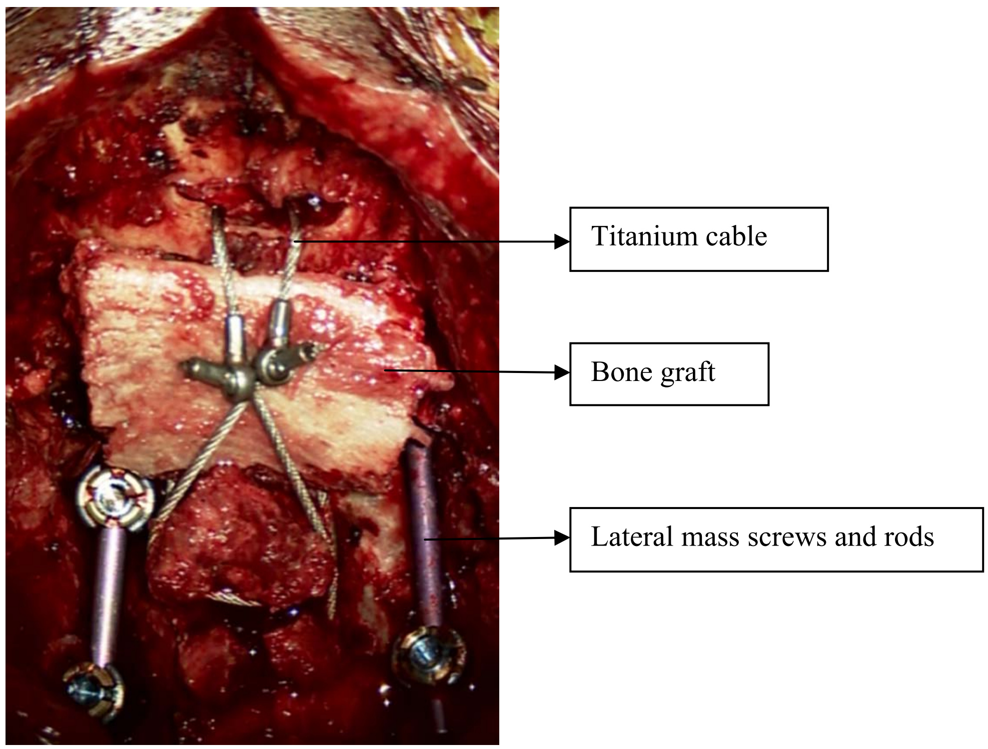
Craniometry:

The various radiological craniometry measures were assessed on preoperative, immediate postoperative and in the follow-up CT scans in the mid sagittal view by 2 neurosurgeons and 1 neuroradiologist. The three observers had a discussion regarding the method of doing these measurements to achieve uniformity. Subsequently the assessments were done independently. The level of the odontoid in relation to the Wackenheim's line, Chamberlain's line, and Mc Rae line were measured. The atlanto-axial distance, narrowest sagittal canal diameter, clival canal angle and the modified omega angle were also measured. The reduction of basilar invagination and the craniovertebral realignment after surgery was determined by comparing the preoperative and postoperative craniometry values. The craniometry measurements of the entire series are tabulated in the appendices 2 and 3.

Surgical technique:

The patients were placed on traction the day prior to surgery to facilitate the reduction of basilar invagination. All patients underwent C1-2 distraction and modified Gallie's posterior fusion. The atlantoaxial facet joints were approached via the pars of C2 and exposed after sectioning the C-2 root bilaterally. The joint capsule was incised and the articular cartilages removed with a micro drill. The joint is distracted bilaterally. Corticocancellous bone is packed into a Globus spacer that is forced into the joints. The size of the spacer is from 6-12 mm and is determined using a trial spacer. A modified Gallie's fusion was done with a braided titanium cable (Medtronic), C1 or occiput to C3 lateral mass screws and rods were inserted. The final construct is shown in the Fig 7.

Figure 7 Final construct of C1-2 distraction and fusion with bone graft



Postoperative management:

Patients are kept in the neurointensive care for 1 day immediately after surgery. They are mobilized the next day with a Philadelphia collar. The postoperative CT scan was done within 1-3 days of surgery. Patients were discharged on the seventh postoperative day after 5 days of intravenous antibiotics. They were advised to wear a cervical collar continuously for a period of 6 months and to review in our outpatient department with a CT and plain X-ray (dynamic view) of craniovertebral junction between 6 months to 1 year of surgery.

Statistical methods:

Both the preoperative and follow-up craniometry findings of three observers were compared and subjected to statistical methods to calculate mean and the significance of reduction of basilar invagination was assessed based on Wilcoxon test using SPSS software version 17 . The mean values of preoperative, follow-up and their difference of the all the three observers were calculated and the inter class correlation was obtained using SPSS software version 17.

Sample size and rationale (41):

The required sample size to show 3 mm reduction as a significant improvement was found to be **20 subjects** after surgery for basilar invagination with 80% power and 5% level of significance. The 3 mm was the minimum reduction of odontoid by Chamberlain's line after realignment surgery for basilar invagination by Goel et al (32).

Formula:

$$n = \frac{(Z_{1-\frac{\alpha}{2}} + Z_{1-\beta})^2}{\Delta^2} + \frac{(Z_{1-\frac{\alpha}{2}})^2}{2}$$

where,

$$\Delta = \frac{(\mu_2 - \mu_1)}{\sigma} \qquad \sigma = \frac{\sigma_1 + \sigma_2}{2}$$

the difference $(\mu_2 - \mu_1) = 3$

RESULTS:

There were 14 males and 6 females aged 14-62 years mean being 32.3 $\pm$ 14.8 years. Two patients had undergone previous surgery for basilar invagination; one underwent C1-2 distraction with bone cement inserted into the C1-2 joint as a spacer done by us while the other patient had a foramen magnum decompression and posterior fusion done elsewhere.

Clinical features:

19 out of 20 patients (95%) presented with features of high cervical myelopathy. 45% of them had neck pain and 35% had torticollis. Fourteen patients were able to walk unaided (70%), four needed support to walk (20%) and two patients were unable to walk even with support (10%). Both spinothalamic and posterior column sensations were affected in 13 patients (65%), posterior column sensations were affected in two patients (10%) and two patients (10%) had only spinothalamic tract involvement. Three patients (15%) had normal sensation. Eight patients (40%) had bowel and bladder symptoms. None had lower cranial nerve symptoms and one patient (5%) had features of respiratory embarrassment which improved postoperatively. He did not require ventilator support postoperatively.

Follow-up:

On follow-up of 7-24 (13.1$\pm$5.23) months, there was significant improvement of all these neurological deficits. There was improvement of neck pain; torticollis was corrected in all these patients. All these patients were able to walk unaided, but five patients (25%) had some residual sensory deficits. The bowel and bladder symptoms improved in all except in one patient (12.5%) where there was worsening of bowel and bladder symptoms. This patient was diagnosed to have worsening of craniometry values on follow up when compared to scan done immediate postoperative period. So he underwent resurgery with replacement of the spacer and screws. On follow up none of these patients had respiratory embarrassment. Overall, 88.8% patients had improvement in their clinical symptoms and signs. The various clinical features are summarized in Table 2.

Table 2 Preoperative symptoms:

Symptoms	**No of patients**	**% of patients**
Neck pain	9	45
Torticollis	7	35
Weakness of limbs		
Able to walk unaided	14	70
Needs support to walk	4	20
Unable to walk	2	10
Sensory deficits		
Normal sensation	3	15
Only spinothalamic tract affected	2	10
Only posterior column affected	2	10
Both spinothalamic and posterior column affected	13	65
Bowel and bladder symptoms	8	40
Respiratory difficulty	1	5
Cranial nerve involvement	0	0

Functional scores:

17 out of 20 patients had a mean follow-up of 13.1$\pm$5.2 months. The mean Nuricks grade improved from 3.2$\pm$1.2 to 2$\pm$1.2 (p value=0.002) postoperatively and modified JOA score improved from 11.1$\pm$3 to 14.7$\pm$2.2 (p value=.000). The details of Nuricks and JOA scores are shown in Tables 3 and 4 respectively.

Table.3 Nurick grade

Follow-up

Preoperative

	0	**1**	**2**	**3**	**4**	**5**
0	**1**					
1						
2	1		**2**			
3	1	1	5	**1**		
4				2	**1**	
5				1	1	

Table 3 shows that 12 patients had at least a grade 1 improvement in their Nuricks score at follow-up while 5 patients (those along the diagonal) had the same Nuricks grade before surgery and at follow-up.

Table. 4 JOA score.

Preoperative \ Follow-up	**7**	**8**	**9**	**10**	**11**	**12**	**13**	**14**	**15**	**16**	**17**	**18**
7						1	1		1			
8												
9					1	1						
10							2					
11										1		
12								2		2		
13									1			
14										1		1
15												
16												1
17												
18												1

Table 4 shows that all the patients had a significant improvement of JOA score at follow-up.

Realignment in immediate postoperative and at follow-up scans:

The craniovertebral junction was realigned and basilar invagination was reduced in 19 of 20 patients (95%) based on the preoperative and postoperative CT scan. All patients in the basilar impression group showed reduction. One patient did not have a satisfactory reduction in the postoperative scan but he had a marked improvement clinically at the last follow-up. One patient had worsening of bowel and bladder symptoms 9 months after surgery but his Nuricks grade improved from 4 to 3 and JOA improved from 7 to 13. The craniometry values of this patient worsened in the follow-up scan when compared to the immediate postoperative scan. He underwent realignment of C1 and C2 and posterior decompression following which his neurological status was stable. 16 out of 17 patients (94%) showed a good bony fusion in the follow-up CT scan. Two patients are yet to come for follow-up, but over a telephonic conversation it was determined they are doing well and attending their work (Nuricks grade of 2). The preoperative, postoperative and the follow-up craniometry values of the patients are tabulated in appendices 2 and 3.

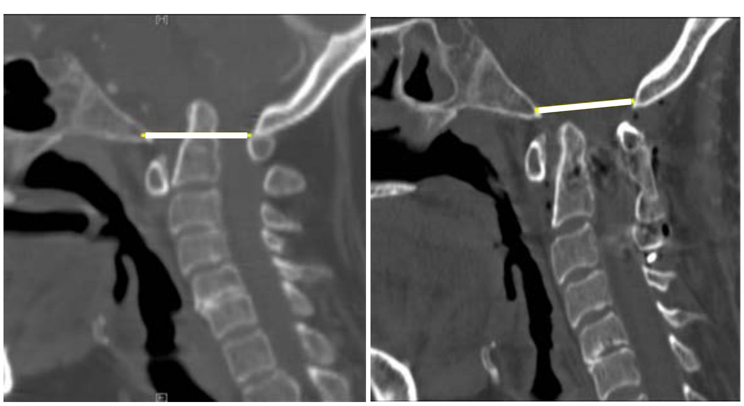
The following (Fig 8) are the illustrated examples of various craniometry findings measured before and after surgery.

Figures 8: Preoperative and postoperative images

1. Chamberlain’s line

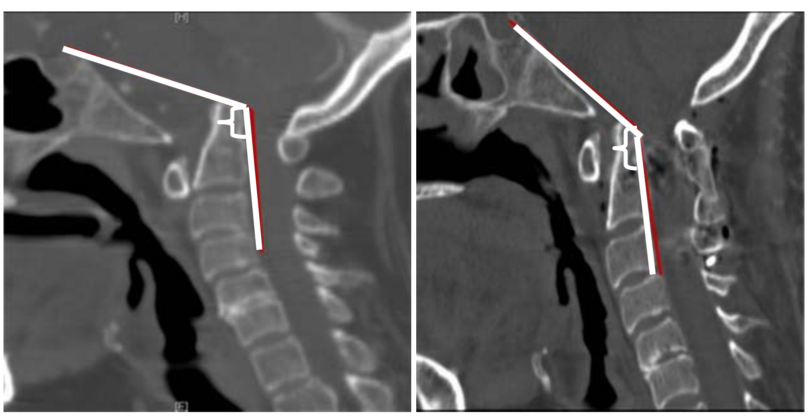
This figures shows excellent realignment of odontoid in relation to various lines before and after surgery

2. Mac Rae’s line



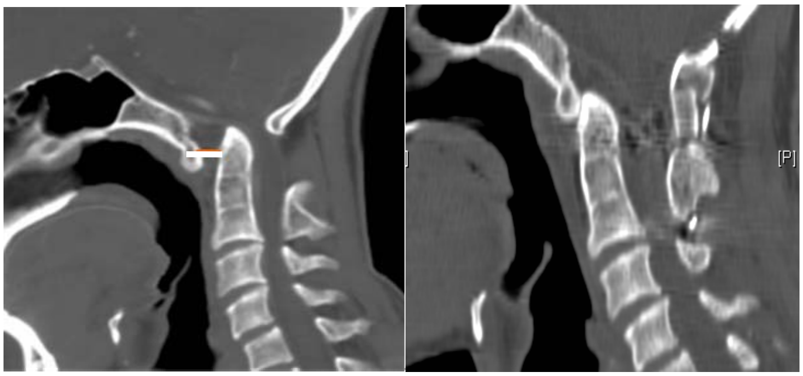
3. Wackenheim's Clival line

4. Clivus canal angle



This figure shows the clival canal angle before and after surgery which was more obtuse after surgery

5. Atlantoaxial distance

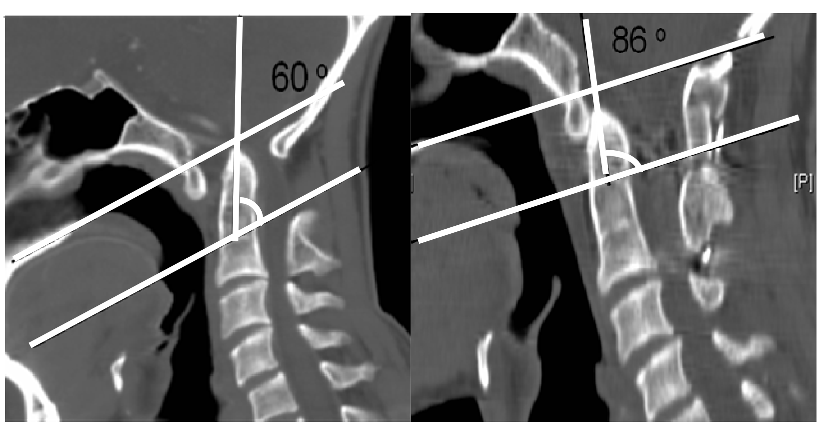


This figure shows the reduction of atlantoaxial distance after surgery

6. Sagittal canal diameter or (Space Available for Cord)

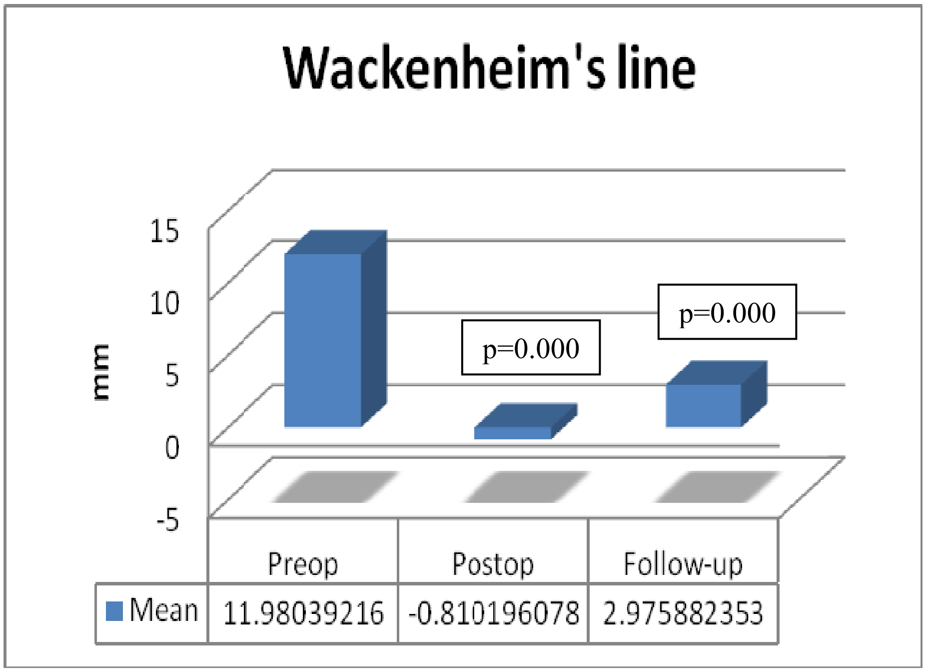
This figure shows the increased space available for cord after surgery.

7. Modified Omega angle



This figure shows change in the modified omega angle before and after surgery

Figure 9: Mean value (of 3 Observers) of the relation of the odontoid to Wackenheim's line in 17 patients.



The mean levels of the odontoid tip above Wackenheim's Clival line on preoperative, postoperative and at follow-up scans are 11.9$\pm$ 5.2, -0.81$\pm$ 5.0and 2.97$\pm$ 5.6 respectively. The minus sign indicates the odontoid tip is below the particular line. The difference of 12.5$\pm$ 4.4mm is achieved due to surgery; however the odontoid has gone back by 3.7$\pm$ 4.8 mm at the follow-up scans. This is depicted in Figure 9.

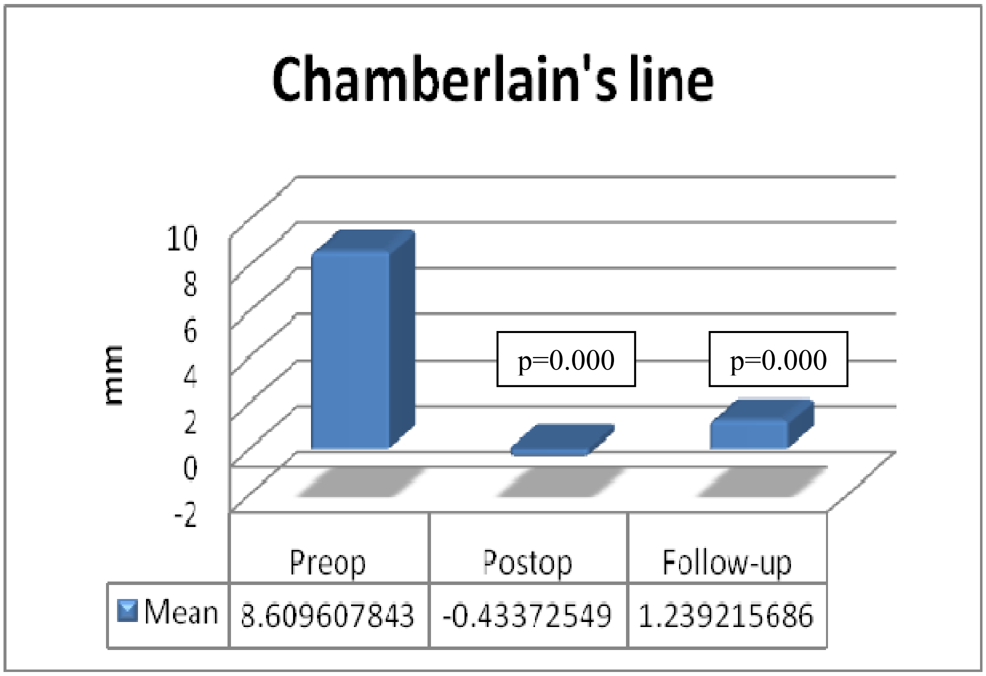
Craniometry measurements:

Table 5 Level of odontoid in relation to Wackenheim's line preoperatively, immediate postop and at follow-up in all patients (mean value of 3 Observers).

Wilcoxon signed-rank test			p=0.000		p=0.000
S. No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	6.7	-8.3	15	-3	9.4
2	14	7.1	7.1	9.2	5
3	11	1.83	9.1	4	6.9
4	16	2.97	13	0.3	16
5	15	0	15	5.1	9.5
6	12	-14	23	2	10
7	7.2	-1.2	8.4	-1	7.8
8	6.6	-2.3	8.9	0.7	5.9
9	**14**	**1.65**	**13**	**16**	**-1.2**
10	14	-2.1	16	2.3	11
11	10	1.33	8.6	4.5	5.4
12	5.1	-0.5	5.6	-0	5.5
13	10	-3.2	14	-3	13
14	27	6.57	20	14	13
15	6.8	-5.4	12	-2	8.4
16	12	0	12	0.6	11
17	16	1.53	14	0	16
18	5.1	-4.4	9.5	N.A	N.A
19	16	4.37	12	N.A	N.A
20	17	6.81	9.9	N.A	N.A

Change I indicates the difference between the level of odontoid before and immediately after surgery. Change II indicates the difference at the follow-up scan compared to preoperative scan. The patient no 9 showed worsening at follow-up who required a resurgery.

Figure 10: Mean value (of 3 Observers) of the relation of the odontoid to Chamberlain's line in 17 patients.



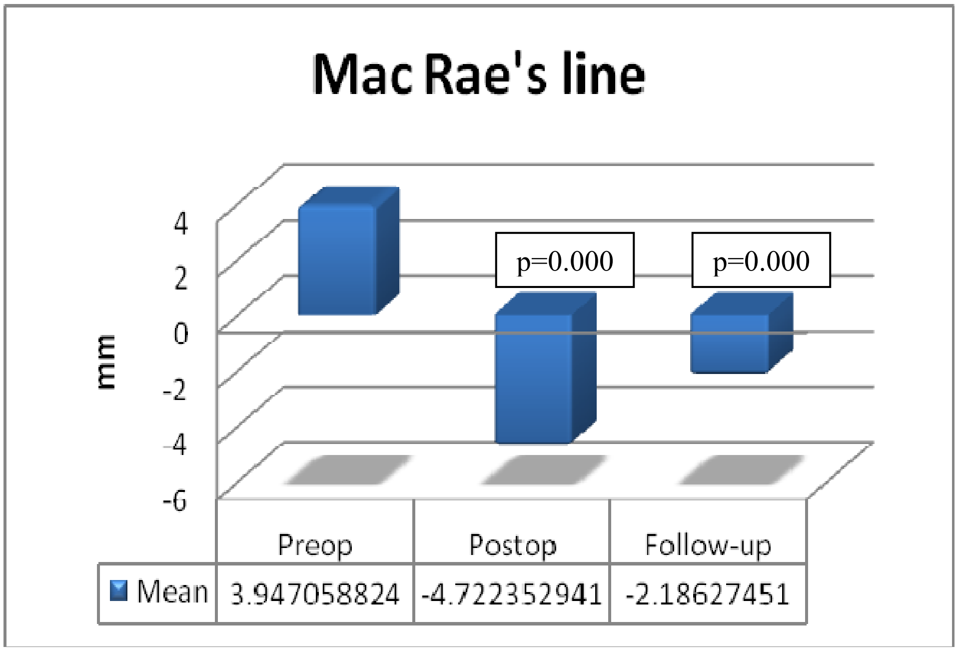
The mean level of odontoid tip in relation to Chamberlain's line at on preoperative, postoperative and at follow-up scans are 8.60$\pm$ 4.3, -0.43$\pm$ 4.0and 1.23$\pm$ 4.28 mm respectively and a mean change of 9 $\pm$ 2.7 mm was achieved due to surgery. There was ascending of odontoid tip by 1.7$\pm$ 3.3mm at follow-up scans. This is depicted in Figure 10.

Table 6 Level of odontoid in relation to Chamberlain's line preoperatively, immediate postop and at follow-up in all patients (mean value of 3 Observers).

	Wilcoxon signed-rank test		p=0.000		p=0.000
S.No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	5.7	-4.4	10	-3	8.2
2	**8.6**	**1.53**	**7.1**	**12**	**-2.9**
3	9.5	2.56	7	0	9.5
4	6.9	1.27	5.6	0.4	6.5
5	14	5.57	8.7	7.4	6.8
6	7.1	-1.7	8.7	3.2	3.9
7	10	-0.3	11	1.8	8.4
8	3.4	-0.8	4.3	-1	4.4
9	-0.1	-8.4	8.4	-4	4.4
10	5.8	-7.1	13	-4	9.6
11	7.8	-3.8	12	0	7.8
12	12	1.1	11	0	14
13	12	3.36	8.8	-2	14
14	17	1.43	15	2.8	14
15	6.2	-1.8	7.9	0	6.2
16	5.2	-2.5	7.7	0	5.3
17	14	6.64	7.8	7.9	6.5
18	14	-10	24	N.A	N.A
19	17	5.69	11	N.A	N.A
20	7.1	1.2	5.9	N.A	N.A

All the patients showed a good reduction at the follow-up scan except patient no 2 who did not have worsening of neurological status at follow-up.

Figure 11: Mean value (of 3 Observers) of the relation of the odontoid to Mac Rae's line in 17 patients.



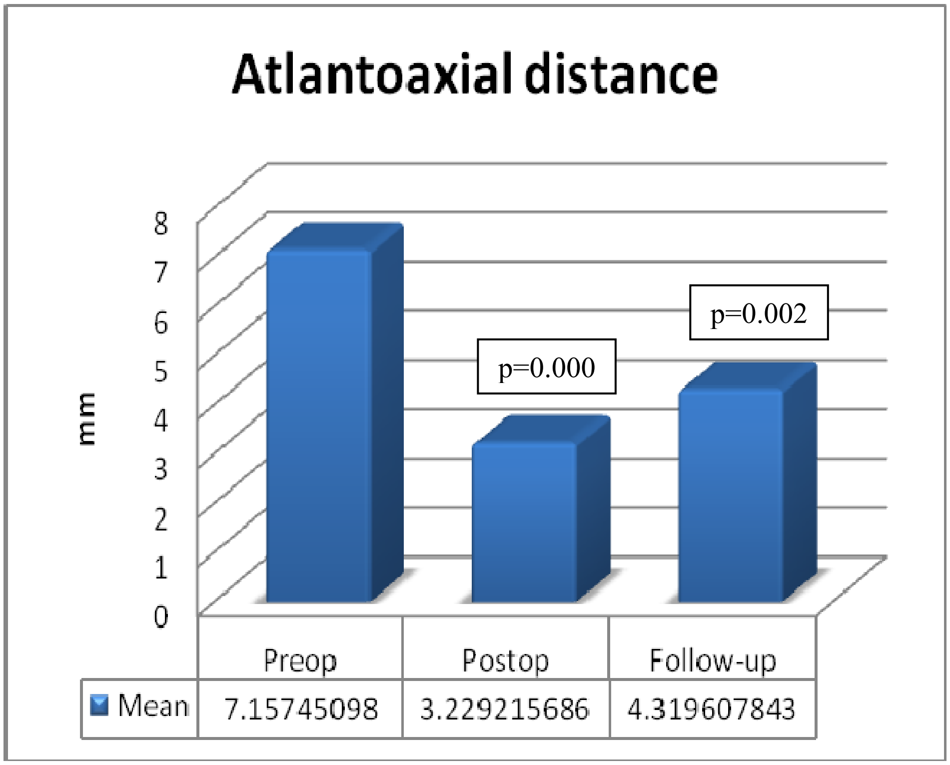
It was seen that the odontoid was 3.9 $\pm$ 5.7 mm above Mac Rae's line preoperatively, while in the immediate postoperative period it was 4.7 mm below this line. The odontoid tip is brought down by 8.6$\pm$ 6.3 mm postoperatively and it is 6.1$\pm$ 3.7 mm below at the follow-up scans when compared to its position in the preoperative scan in relation to Mac Rae's line. This is depicted in Figure 11.

Table 7 shows the level of odontoid in relation to Mac Rae's line in all patients (mean value of 3 Observers)

Wilcoxon signed-rank test			p=0.000		p=0.000
S.No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	-0.2	-11	11	-3.5	3.3
2	**3.06**	**-3.3**	**6.4**	**3.1**	**-0**
3	10.3	-0.8	11	3.33	6.9
4	2.2	-2.6	4.8	-2.4	4.6
5	7.13	-1.9	9.1	0.67	6.5
6	0.76	-10	11	-5.9	6.7
7	3.03	-4.5	7.6	-1.5	4.5
8	0.2	-7.2	7.4	-9.6	9.8
9	-0.4	-9.8	9.3	-3.7	3.3
10	2.57	-7.4	10	-3.6	6.2
11	1.93	-4.6	6.5	-3.6	5.6
12	5.6	-2.5	5.5	-6.7	12
13	5.33	-4.1	9.4	-4.7	10
14	12.6	-2.7	12	0	13
15	3.83	-4.3	8.1	0.57	3.3
16	1.27	-7.3	8.5	-5.4	6.7
17	7.97	4.51	8.5	5.87	2.1
18	5.25	-2.7	7.9	N.A	N.A
19	11.7	-0.1	12	N.A	N.A
20	6.73	1.2	5.5	N.A	N.A

All the patients showed good reduction of odontoid in relation to Mac Rae's line except patient no 2 who did not require a intervention because of stable neurological status.

Figure 12: Mean value (of 3 Observers) of the atlantoaxial distance in 17 patients.



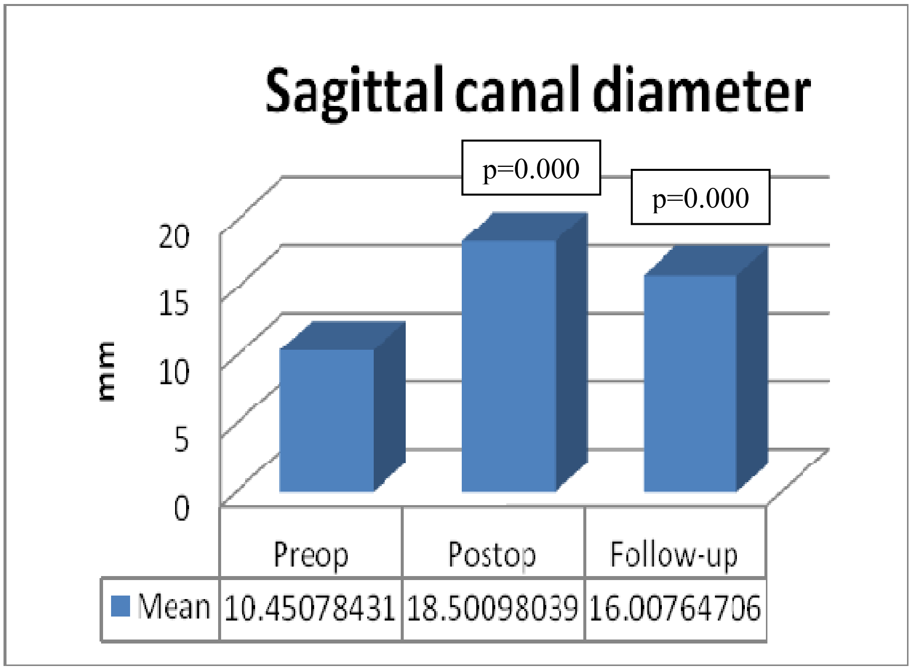
It was shown that the mean value of atlantoaxial distance before surgery was 7.1 $\pm$ 2.4 and it was 3.2 $\pm$ 2.7 mm postoperatively and it is increased to 4.3 $\pm$ 3.2 mm at follow-up. The difference of 2.83$\pm$ 2.6 mm is achieved due to realignment surgery at the follow-up scans. This is depicted in Figure 12.

Table 8 shows the atlantoaxial distance in all patients (mean value of 3 Observers)

	Wilcoxon signed-rank test		p=0.000		p=0.002
S.No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	6.13	5.33	0.8	5.1	1
2	**11.5**	**10.3**	**1.2**	**13.3**	**-2**
3	6.21	1.1	5.1	3.03	3.2
4	12	6.2	5.8	4.77	7.2
5	8.63	1.13	7.5	3.7	4.9
6	9.13	2.77	6.4	6.87	2.3
7	5.93	0.83	5.1	1.87	4.1
8	5.33	0.9	4.4	0	5.3
9	**4.95**	**3.3**	**1.6**	**8.08**	**-3**
10	11.4	5.71	5.7	7.35	4.1
11	4.53	1.95	2.6	2.4	2.1
12	6.24	4.95	1.3	4.67	1.6
13	6.48	2.73	3.8	2.37	4.1
14	3.87	5.67	-2	3.87	0
15	7.33	0.67	6.7	2.19	5.1
16	6.37	1.07	5.3	3.2	3.2
17	5.65	0.33	5.3	0.73	4.9
18	6.47	2.3	4.2	N.A	N.A
19	3.5	3.2	0.3	N.A	N.A
20	12.3	11	1.3	N.A	N.A

Patient no. 2 and 9 showed increase in the atlantoaxial distance at the follow-up scan. Rest of the patients showed a decrease in the atlantoaxial distance after surgery and at follow-up. As already mentioned patient no 9 underwent a resurgery whereas the patient no 2 was stable neurologically at the follow-up and he did not require any intervention.

Figure 13: Mean value (of 3 Observers) of the sagittal canal diameter in 17 patients.



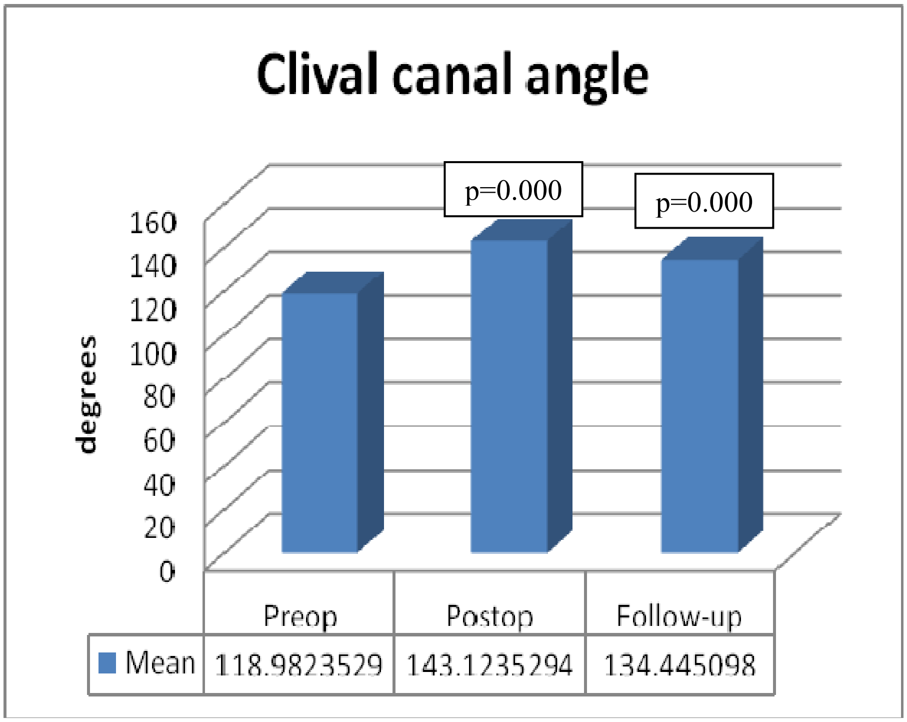
It was shown that the mean value of sagittal canal diameter before surgery was 10.5 $\pm$ 2.9 and it was increased to 18.5 $\pm$ 4 mm postoperatively and it is decreased to 16 $\pm$ 4 mm at follow-up. The difference in the increase in the size of the canal was 5.55$\pm$ 3.6 mm at the follow-up. This is depicted in Figure 13.

Table 9 shows the sagittal canal diameter in all patients (mean value of 3 Observers)

	Wilcoxon signed-rank test		p=0.000		p=0.000
S.No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	8.5	18.8	10	18	9.3
2	12	16.4	4.6	13	1.7
3	8.2	16.9	8.7	18	9.5
4	7.6	14.7	7.1	16	8.6
5	9.5	19.5	10	16	7
6	**10**	**21.1**	**11**	**9.3**	**-0.9**
7	11	15.7	4.9	13	2.6
8	12	16.8	4.7	18	6
9	6.4	13	6.6	7.8	1.4
10	12	22	10	17	5.2
11	17	23.3	6.3	22	5.1
12	14	20.7	6.3	17	2.6
13	7.8	13.7	5.9	13	5.7
14	9.7	12.9	3.2	11	1.8
15	7.5	18.6	11	17	9.5
16	15	26.2	11	22	6.9
17	9.1	24.1	15	22	13
18	12	18.6	6.2	N.A	N.A
19	12	16.2	4.1	N.A	N.A
20	5.1	13.8	8.7	N.A	N.A

All the patients had a significant increase in the size of sagittal canal diameter which is the space available for the cord except the patient no 6 who had worsening of the space at the follow-up scan. She had a very good neurological recovery at the last follow-up.

Figure 14: Mean value (of 3 Observers) of the Clival canal angle in 17 patients.



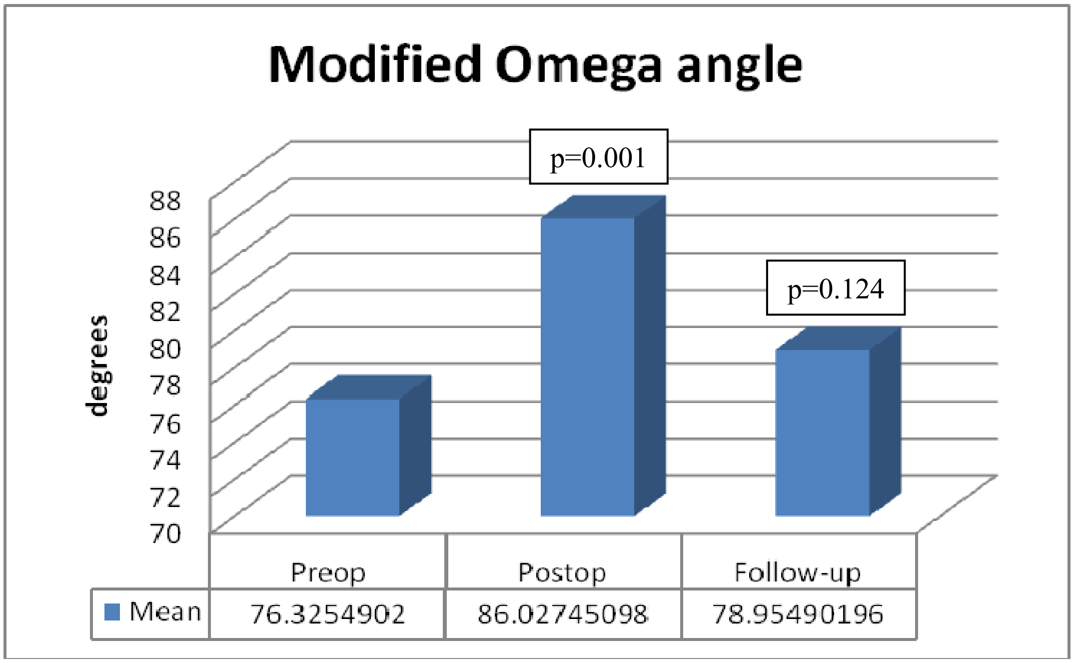
It was shown that the mean value of clival canal angle before surgery was 119 $\pm$ 17 degrees and it was increased to 143.1 $\pm$ 12.8 degrees postoperatively and it is decreased to 134 $\pm$ 18 degrees at the follow-up. The change in the angle at the follow-up scan, 15.4 $\pm$ 10.2 degrees is achieved due to surgery. This is depicted in the Figure 14

Table 10 shows the clival canal angle in all patients (mean value of 3 Observers)

Wilcoxon signed-rank test			p=0.000		p=0.000
S.No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	107	146	40	139	33
2	105	112	6.6	106	0.3
3	114	141	27	143	29
4	113	148	35	142	30
5	136	148	12	143	6.9
6	101	142	42	108	7.7
7	117	134	17	128	11
8	150	166	16	172	22
9	79.7	121	41	98	18
10	129	152	23	132	3.1
11	124	151	27	143	19
12	123	146	23	140	18
13	134	156	22	153	19
14	108	133	25	122	14
15	114	148	34	139	37
16	141	147	6.6	141	0.2
17	128	141	13	137	8.6
18	123	130	6.8	N.A	N.A
19	103	111	8	N.A	N.A
20	95.4	125	29	N.A	N.A

All the patients showed increase in the clival canal angle which is a consistent finding among all the craniometry measured. This is due to downward as well as anterior realignment of odontoid due to surgery.

Figure 15: Mean value (of 3 Observers) of the Modified Omega canal angle in 17 patients.



It was shown that the mean value of Modified Omega angle before surgery was 76.3 $\pm$ 16 degrees and it was increased to 86 $\pm$ 11.8 degrees postoperatively and it is decreased to 79 $\pm$ 12.1 degrees at follow-up. This is depicted in Figure 15.

Table 11 shows the modified omega angle in all patients (mean value of 3 Observers)

Wilcoxon signed-rank test			p=0.001		p=0.124
S.No	Preop	Immediate Postop	Change 1	Follow-up	Change 2
1	60	81.7	21	85	25
2	**60**	**60.3**	**0**	**55**	**-4.9**
3	79	86.9	8.1	80	1.5
4	70	91.2	21	85	15
5	**100**	**92.1**	**-8**	**88**	**-12**
6	64	89.1	25	64	0.3
7	77	87.6	11	78	1.6
8	**96**	**104**	**8.6**	**74**	**-22**
9	37	59.1	22	53	15
10	83	94.3	12	89	5.9
11	74	85.9	12	77	3.6
12	84	91.6	7.2	89	4.2
13	77	74.7	-2.1	82	5.5
14	82	88.5	6.9	82	0.3
15	66	88	22	73	7
16	**95**	**89.2**	**-6.1**	**92**	**-3.5**
17	94	97.9	4.1	96	2.7
18	80	88	7.9	N.A	N.A
19	77	81.3	4.1	N.A	N.A
20	56	79.7	24	N.A	N.A

Patient no. 2, 5, 8, and 16 showed minimal decrease in the angle at the follow-up scans. The patients who had an increase in the angle at the follow-up scans are shown to have no significance. This change in the angle is very inconsistent among the craniometry measured.

Inter class correlation (ICC):

The inter class correlation between all the three observers was calculated based on the preoperative and follow-up craniometry data are shown in Table 12.

Table 12

ICC between observers based on preoperative and the follow-up craniometry findings:

Variables	ICC	ICC
Wackenheim's Clival line	.849	.881
Chamberlain's line	.885	.889
Mac Rae's line	.718	.787
Atlantoaxialdistance	**.627**	.922
Space Available for Cord	.825	.826
Clival canal angle	.853	.941
Modified omega angle	.859	**.308**

The atlantoaxial distance had a fair to good agreement between observers among the preoperative variables and the rest of the variables had an excellent agreement at follow-up except modified omega angle at follow-up which was very poor.

DISCUSSION:

Craniovertebral anomalies are frequently found in the Indian subcontinent particularly in Uttar Pradesh, Bihar, Rajasthan and parts of Gujarat (4). The surgical management of congenital craniovertebral anomalies is complex due to the relative difficulty in accessing the region, critical relationships of neurovascular structures and the biomechanical issues involved.

Morbidity of transoral surgery:

The transoro -pharyngeal exposure is the most common approach for ventral decompression. Jain et al (15) studied the surgical outcome of 74 patients, who underwent transoral decompression. The major morbidity in their study was pharyngeal wound sepsis leading to dehiscence (20.3%) and hemorrhage (4%), velopharyngeal insufficiency (8.1%), CSF leak (6.7%) and inadequate decompression (6.7%). Neurological deterioration occurred transiently in 17 (22.9%) and was sustained in 7 (9.4%) patients. Naderi et al (23) reported further cranial settling in two patients whom underwent transoral decompression and occipitocervical fusion. The complications of the approach include contamination by normal oral flora (17, 18) , dehiscence of the posterior pharynx, alteration in phonation secondary to effects of surgery on the pharynx (19, 20), tongue edema (21), the potential need for prolonged intubation (21), and the requirement of avoiding oral intake to allow the pharyngeal closure to heal (20). Other complications include vertical occipitoatlantal subluxation with vertebral artery occlusion and brain stem stroke (22). CSF leaks encountered during the course of a transoral surgery have potentially devastating consequences. Meningitis caused by oral bacteria invading the CSF and death have been reported with this technique (18). Menezes et al (16) had 2 unrelated deaths, one due to a myocardial infarction and the other was due to septicemia among 72 patients of a transoral series. Goel had a morbidity of 1% in his transoral series of 99 (10).

Pathogenesis of basilar invagination:

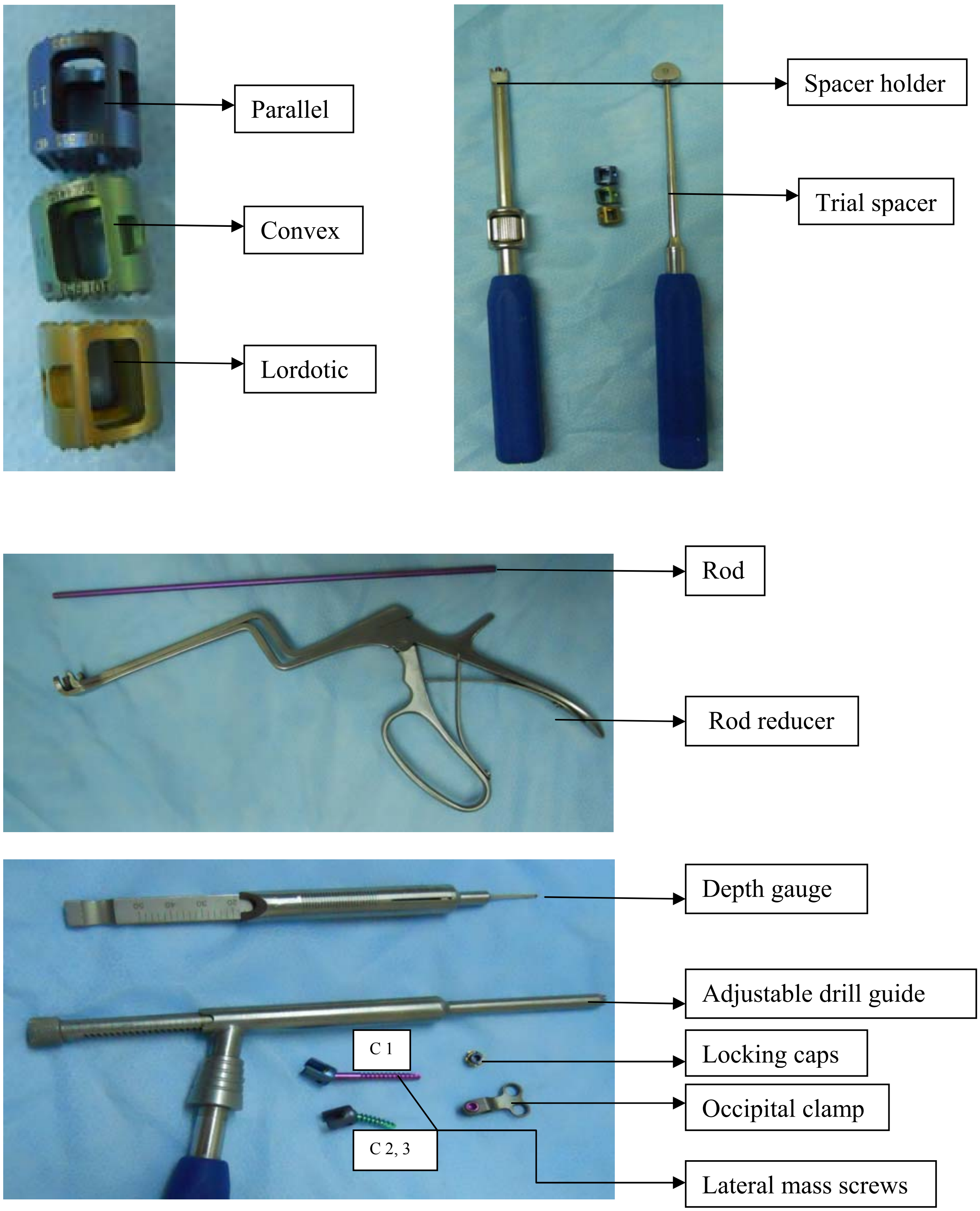
Goel et al speculated that basilar invagination is probably secondary to “slippage” of the atlas over the axis (31). This slippage can be severe enough to cause spondyloptosis of the atlas over the axis. These authors reported a novel surgical technique in which the cranially migrated odontoid is brought down to its normal relationship with the atlas and fixed in position. In their technique a spacer is connected to a stainless steel plate and is used for distracting the C1-2 joint. The mean reduction of the odontoid achieved was 7.5, 8.7, and 6.2 mm corresponding to Wackenheim’s, Chamberlain’s and Mac Rae’s line respectively (31). All their patients improved postoperatively and there were no complications reported in their series. They considered distraction and fusion surgery superior to other treatment options for basilar invagination as it is deals directly with the pathogenesis of basilar invagination and has less morbidity.

Basis for our study:

We did this study using Goel’s technique, however modified the instrumentation. We designed titanium spacers (lordotic, parallel and convex) as shown in Figure 16 that were instrumented into the C1-2 joint and placed separate lateral mass screws into the C1 and C2 or C3. In addition we performed a modified Gallie’s fusion using a titanium cable and iliac bone graft. We present the clinical and the radiological findings in 20 patients with basilar invagination type A (Goel et al) who underwent distraction surgery. The craniometry findings were performed by three observers, 2 neurosurgeons and 1 neuroradiologist to avoid bias and the interobserver correlation coefficient was calculated.

Figure 16 Implants and instruments used in craniovertebral junction realignment surgery

Types of spacers



Description of surgical technique:

The patient is positioned prone on traction with 5- 8 kg and the head end of the bed elevated. A midline skin incision is made from the external occipital protuberance to C4. The foramen magnum and laminae of C2-4 are exposed. Both C1-2 joints are exposed along the pedicles of C2. The C2 roots on both sides are cut and the C1/2 joints opened bilaterally. Using a small diamond drill the joint cartilages are drilled. A trial spacer is initially introduced (Figure 16) into the joint space to determine the size and type of the spacer needed to distract the joints as well as to correct the head tilt to a certain extent. A final titanium spacer with bone chips is inserted into the C1/2 joints. In patients with occipitalized C 1 arch foramen magnum decompression is done by drilling off the occipitalized C1, using a high speed drill the occiput is drilled in the midline to expose the dura about 1 cm above the foramen magnum to create an artificial C1. A braided titanium wire is passed through the occipital hole and looped around the C2 spine around a groove made with a drill. The lateral mass of C1 and C2 or C3 is drilled with a adjustable drill guide (shown in Figure 16) which measures the length of screws. After drilling, the length of the screw needed is assessed by passing depth gauge (shown in Figure 16). The appropriate lateral mass screws are inserted bilaterally. The half thread shoulder screws (shown in Figure 16) are used in C1 lateral mass and full threaded screws (shown in Figure 16) are used in the C2 or C3 lateral mass. Rods (shown in Figure 16) are contoured and fixed from the C1 to C3 lateral mass screws using locking caps (shown in Figure 16). Occasionally, particularly in children occiput clamps instead of C1 lateral mass screws is used (shown in Figure 16). The modified Gallie's fusion is done as described earlier. The wounds are irrigated and closed in layers with drains. The final construct is shown in Figure 7.

Clinical outcome:

On follow-up of 7-24 (13.1±5.23) months, there was significant improvement in neurological deficits. There was improvement of neck pain; torticollis was corrected in all these patients. All patients were able to walk unaided but there were five (25%) who had some sensory deficits. In general 88.8% of our patients had improvement in their clinical symptoms and signs. There are 12 patients who had improvement by at least 1 grade at follow-up. Three patients (two patients who needed support to walk and one was bed bound) are able to walk without support at follow-up. These functional scores are neither assessed nor compared before surgery and at follow up in other studies (4). These results are comparable to the study done by Goel et al (4). In their series, neck pain was observed in 77% and torticollis in 41% of their patients and all of them improved postoperatively. The preoperative and follow-up Nuricks grade and JOA score are plotted in table 3.

Radiological outcome:

Immediate:

The craniovertebral junction was realigned and basilar invagination was reduced in 19 out of 20 patients (95%) with significant improvement in the craniometry measurements. Restoration of craniovertebral realignment is easier in patients with rheumatoid arthritis as it is due to the pathology of ligaments rather than the bone itself as mentioned by Goel et al (33). One patient did not have satisfactory reduction in the immediate postoperative scan however since he improved he was followed up and was found to be stable at follow-up.

Follow up radiology:

During the follow-up period (13.1±5.23 months) the craniometry findings in the follow-up scans showed a slight change as compared to the immediate postop CT scan indicating settling. When early CT scan are done 1-3 days after surgery most patients may not be fully ambulated and this may not really reflect the true position of the odontoid in relation to all the lines and it seems to reduce on follow-up. However the importance of the immediate postoperative CT is to show good alignment before sending the patient home. Patients tend to have some degree of cranial settling in the follow-up scans when compared to the immediate post operative scans but bony fusion occurred in most of them (94%). One patient had significant worsening of the craniometry values when compared to the immediate postoperative scan along with worsening symptoms. He underwent realignment of C1 and C2 and posterior decompression following which his neurological status was stable.

The mean preoperative and follow-up craniometry subjected to statistical methods was shown statistically significant by all the observers except the modified omega angle which was not statistically significant. The modified omega angle corresponds to the anteroposterior tilt of the odontoid and postoperatively this angle did not change enough to cause statistical significance.

Wackenheim's line:

The mean levels of the odontoid tip above Wackenheim's Clival line on preoperative, postoperative and at follow-up scans are 11.9± 5.2, -0.81± 5.0and 2.97± 5.6 respectively. The minus sign indicates the odontoid tip is below the particular line. The difference of 9 ± 4.3 mm is achieved due to surgery. This reduction is better than that achieved by Goel et al who found reduction of 7.5 mm (31).

Chamberlain's line:

The mean reduction of odontoid tip in relation to Chamberlain's line is 7.46 $\pm$ 4.2 mm which is comparable to the results by Goel et al which is 8.7 mm.

Mac Rae's line:

The mean reduction of the odontoid in relation to Mac Rae's line is 6.1 $\pm$ 3.7 which is comparable to achieved by Goel et al which is 6.2$\pm$ mm corresponding to Mac Rae's line (31).

Atlantoaxial distance:

The mean reduction of atlantoaxial distance achieved due to realignment surgery is 2.83$\pm$ 2.6 mm which is 4.7 mm mean by Goel et al (31).

Space available for cord or Sagittal canal diameter:

The space available for the cord is increased to 16 $\pm$ 4.1 mm due to surgery at the follow-up scans. The difference in the increase in the size of the canal was 5.55$\pm$ 3.6 mm. This extensive increase in the size of the canal is directly proportional to the excellent clinical improvement noticed in these patients.

Clival canal angle:

The mean Clival canal angle at follow up is 134.44 $\pm$ 18 degrees which is the normal range (130-150 degrees). The change in the angle due to surgery is 15.5$\pm$ 10.2 degrees. This indicates the downward as well as anterior realignment of odontoid due to surgery.

Modified omega angle:

This angle was changed by 2.62$\pm$ 5.85 degrees due to surgery which is not statistically significant (p value=0.12). This angle change indicates the anterior displacement of the odontoid tip due to surgery. Goel et al (31) achieved mean change of 15.7 degrees in his series which is better than us.

Interoberver correlation coefficient:

ICC analysis showed an excellent agreement between observers in measuring various craniometries. The preoperative atlantoaxial distance had a agreement of only 0.627 and at the follow-up scan is .922. The reason for this excellent agreement is the atlantoaxial distance value is close to zero in the follow-up scans. The follow-up modified omega had a value of .308 which is a poor agreement. This poor agreement is probably because in some of the postoperative scans the anterior end of the hard palate is not adequately covered to draw the tangential line which passes through the base of C2 body.

Complications:

One of our patients had a vertebral artery injury during the surgery. This patient presented with features of cervical cord compression, Nuricks grade of 3 and modified JOA score of 13/18. He was diagnosed to have basilar invagination and underwent foramen magnum decompression and occipitocervical fusion elsewhere and his neurological status continued to worsen after his first surgery. He underwent C1-2 distraction and occipitocervical fusion and had a vertebral artery injury during surgery. His Nuricks worsened to grade 5 and JOA to 7/18 immediately after surgery and was discharged with a bed bound status and on Ryle's tube feed. This patient was lost to follow-up.

Reoperation:

In our series, one out of 20 patients underwent re-alignment after first surgery. He presented with Nuricks grade of 4 and JOA score of 7/15. Immediate postoperative period his Nuricks grade improved to 3. On follow up after 1 year his Nuricks grade improved to 3 and JOA to 13/18. Although there was improvement of his tightness and weakness of limbs and sensory deficits he continued to have worsening of bowel and bladder symptoms. There was worsening of craniometry findings when compared to the scan done in the immediate postoperative period as shown in Fig 17.

Figure 17 Midsagittal CT of the patient who underwent reoperation. There was definite worsening of the craniometry at follow-up scan when compared to the immediate postoperative scan

Preoperative | Immediate postoperative | Follow-up

A | B | C

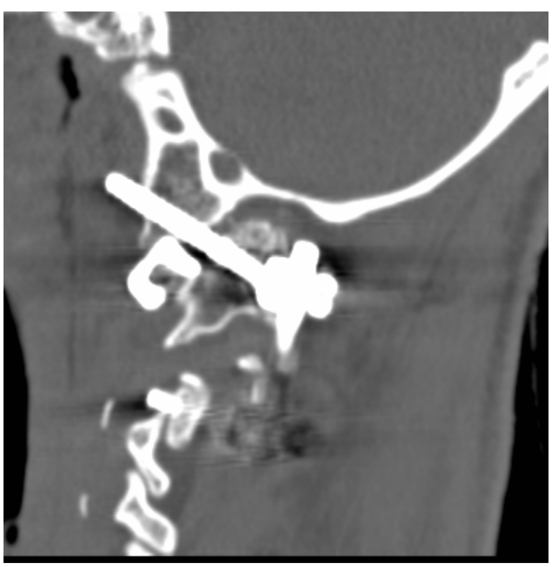
His neurological status stabilized after the second surgery. This is probably due to the implant related complications which is shown in figure 18 A. The spacer was placed more anteriorly in the left C1-2 joint which probably caused the reinvagination.

Figure 18

A

Immediate postoperative scan after the 2 nd surgery showing realignment of craniovertebral junction and widening of the space available for the cord.

B



Immediate postoperative scan after I st surgery showing slippage of the spacer anteriorly.

Realignment surgery in comparison with transoral odontoidectomy:

We agree with Goel et al that the results of distraction surgeries are superior to the transoral surgeries. Although Goel et al (31) report 100% success rate with no clinical deterioration or implant related complications in their series, we report a 94.1 % of clinical improvement at the last follow-up, 5% is our morbidity rate due to a vertebral artery injury and 5% is implant related complications. There was no mortality. Most transoral series report an improvement ranging from 36.8% (27) to 100% (10, 16 and 22), morbidity ranging from 1% (10) to 22.9% (15) and mortality ranging from 2.6% (27) to 13.3% (7).

CONCLUSION:

Craniovertebral realignment provides excellent neural decompression and clinical outcome in patients with basilar invagination. The C 1-2 distraction with placement of a spacer into the C1-2 joints bilaterally helps reduce the C1 2 slippage and basilar invagination. Thus the surgery deals directly deals with the pathology. It is a technically demanding surgery and accurate placement of the implants plays a vital role in the reduction of basilar invagination. Previous craniovertebral surgery with risk of damage to the vertebral artery is probably a contraindication to the procedure. The reduction of basilar invagination in the early postoperative scan and the position of the spacer may be a good predictor for long term results even though our sample size is small. Although good bony fusion is seen by 6 months, short term follow-up indicates some settling of graft and spacer. Therefore longer follow-up is mandatory before considering C1 2 distraction surgery as a gold standard treatment for basilar invagination.

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Appendix -1

C1-2 DISTRACTION AND FUSION PROFORMA

Name- Hospital no-

Age- Sex Occupation

Address-

Nuricks grade-

JOA score-

CLINICAL FEATURES:

Neck pain: Present / absent

Torticollis/ restricted neck movements: Present / Absent.

Weakness of limbs:

Able to walk unaided/ Needs support to walk/ Unable to walk with support

Sensation:

Normal sensation/Spinothalamic function affected/ Posterior column affected/ Both spinothalamic and posterior column affected.

Bowel/bladder symptoms- Present / Absent.

Cranial nerve involvement – Present/ Absent

Respiratory embarrassment- Present/ Absent.

History of trauma- Present/Absent

History suggestive of infection (TB) -Present/ Absent

History suggestive of inflammation (Rheumatoid arthritis) - Present/ Absent

History of previous surgery

Preoperative traction applied- Yes/ No

Improvement of neck pain- Yes/ no

Improvement of weakness of limbs Yes/ no

Improvement of sensation- Yes/ no

Improvement of respiratory embarrassment - Yes/ no

OPERATION DONE

C1-2 DISTRACTION WITH SPACER AND POSTERIOR FUSION WITH 1--3 /1-2 LATERAL MASS SCREW AND ROD FIXATION

Size of spacer:

Right	Left

Postoperative clinical findings;

Improvement of neck pain- Yes/ no

Improvement of weakness of limbs Yes/ no

Improvement of sensation- Yes/ no

Improvement of respiratory embarrassment - Yes/ no

Radiographic findings:

MRI findings

Evidence of cord compression. Yes/no

Hyper intensity changes in cord Yes/no

Saggital canal diameter $\geq$ 20 $<$ 20 mm

Chiari malformation Present/ Abssent

CT FINDINGS

CT findings	Preoperative	Postoperative
Wackenheim's line	Above /below mm	Above/below mm
Chamberlain's line	mm	mm
Mc Rae's line	Above/below mm	Above/below mm
Atlantoaxial distance	mm	mm
Clival canal angle		
Omega angle.		
Sagittal canal diameter		
Other deformities		

C1-2 Distraction follow up proforma

Follow up in months ---

Symptoms

Neck pain – Improved/static / worsened

Weakness of limbs – Improved/static / worsened

Sensory symptoms – Improved/static / worsened

Bowel or bladder symptoms- – Improved/static / worsened

Respiratory embarrassment – Improved/static / worsened

New symptoms present/ absent

Nuricks grade-

JOA score-

CT FINDINGS

CT findings	preop	-- months follow up
Wackenheim’s line	Above /below mm	Above/below mm
Chamberlain’s line	mm	mm
Mc Rae’s line	Above/below mm	Above/below mm
Atlantoaxial distance	mm	mm
Clival canal angle		
Omega angle		
Sagittal canal diameter		

Appendix-2

Preoperative and postoperative CT scan data measured by Observer 1

S.no	wlpre1	wlpos1	change	clpre1	clpos1	change	mlpre1	mlpos1	change	aadpre1	aadpos	change	ccapre1	ccapos	change	scpre1	scapos1	change	ompre1	ompos1	change
1	4.65	-8.73	13.38	4.65	-2	6.65	-0.7	-9.5	8.8	5.9	5.9	0	109.1	141.9	32.8	8.8	19.4	10.6	56.8	74.4	17.6
2	17.7	6.5	11.2	10.6	0	10.6	8.2	-4.8	13	17.4	10	7.4	88.9	112.3	23.4	8.1	20.2	12.1	52.5	64.6	12.1
3	11.36	0	11.36	8.45	0	8.45	9.9	-2	11.9	5.24	0.8	4.44	119.3	147.9	28.6	9.2	18	8.8	78.6	87.9	9.3
4	15.1	0	15.1	5.9	0	5.9	0	-4.4	4.4	13	7.1	5.9	103.3	142.4	39.1	9	15.9	6.9	70.6	90.3	19.7
5	6	-5	11	14	-10	24	5	-3	8	7	2	5	120	130.4	10.4	12	19	7	79	89.4	10.4
6	12.4	0	12.4	16.2	6.4	9.8	9.1	0	9.1	9.6	0	9.6	133.3	146.1	12.8	10.2	19.8	9.6	96.5	97.4	0.9
7	2.36	-13.7	16.1	7.88	-1.5	9.38	5.58	0	5.58	12.6	3.6	9	100.5	139.9	39.4	12.44	22.29	9.85	63.9	90.7	26.8
8	6.9	0	6.9	12.2	0	12.2	2	-6.5	8.5	6.9	0	6.9	107.8	131.4	13.6	11.5	19.6	8.1	77.9	93.7	15.8
9	7.96	0	7.96	4.95	0	4.95	5.6	-2	7.6	4.5	0	4.5	150.3	165.6	15.3	12.4	17.2	4.8	98.7	108.1	9.4
10	13.2	7.2	6	0	-10.6	10.6	0	-10.5	10.54	4.04	3.2	0.84	72.8	116.5	43.7	6.8	14.56	7.76	34.9	50.5	15.6
11	16.6	0	16.6	5.6	-4.8	10.4	1.7	-7.3	9	12.46	6.1	6.36	129.4	154.6	25.2	13.2	31.3	18.1	81.6	97.2	15.6
12	9.3	0	9.3	9.4	-5.9	15.3	3.7	-5.4	9.1	5.4	1	4.4	123.3	147.3	24	17	22.4	5.4	65.3	88.4	23.1
13	4	0	4	13.7	0	13.7	4.5	-7	11.5	6.5	5.9	0.6	124.8	150	25.2	14	20	6	85	89	4
14	10.7	-2.8	13.5	12.7	4	8.7	5	-1.5	6.5	6.3	3.3	3	132.6	156.4	23.8	8	13.6	5.6	69.9	74.7	4.8
15	14.7	2.2	12.5	13.63	6.88	6.75	9.65	1.4	8.25	4	1.4	2.6	99.2	109.6	10.4	10.5	20	9.5	73.2	83.2	10
16	26.5	5	21.5	17.5	0	17.5	13.9	-2.7	16.6	5	5	0	105.3	133.2	27.9	10.3	13.1	2.8	79.3	88.6	9.3
17	6.6	-1.2	7.8	6.6	0	6.6	4.6	-4.7	9.3	8.8	0	8.8	117	149	32	9	21.9	12.9	65.7	99.6	33.9
18	9.5	0	9.5	5.8	0	5.8	0	-8.8	8.8	7.7	0	7.7	137.6	147.2	9.6	15.9	33	17.1	90.7	90.3	-0.4
19	12.9	3	9.9	4.2	0	4.2	4.3	0	4.3	13.8	12	1.8	94.3	124.2	29.9	5.4	14.3	8.9	60.3	86.5	26.2
20	15.4	0	15.4	11.14	5.7	5.44	8.2	2.3	5.9	4.9	0	4.9	133	140.9	7.9	8.7	21.4	12.7	92.4	98.2	5.8

Preoperative and postoperative CT scan data measured by Observer 2

S.no	wlpre2	wl pos2	change	clpre2	clpos2	change	mlpre2	mlpos2	change	aadpre2	aadpos2	change	ccapre2	ccapos2	change	sacpre2	sacpos2	change	ompre2	ompos2	change
1	6.04	-5.4	11.44	6.5	-3.6	10.1	0	-11.7	11.7	4.3	4.09	0.21	95.3	143.1	47.8	7.14	20.8	13.66	62.8	82.3	19.5
2	12.54	7.01	5.53	8.82	4.6	4.22	0.98	0	0.98	7.84	9.56	-1.72	106	106.9	0.9	13.84	13.76	-0.08	63.6	57.2	-6.4
3	10.38	2.2	8.18	11.07	3.97	7.1	11.53	3.97	7.56	6.1	1	5.1	105.9	134.1	28.2	7.69	17.1	9.41	77.3	85.9	8.6
4	15	3.3	11.7	9.37	3.8	5.57	3.7	-1	4.7	11.25	5.9	5.35	111.6	147.3	35.7	5.6	14	8.4	71.2	87.4	16.2
5	2.56	-3.46	6.02	15.24	-9.24	24.48	6.36	-2.1	8.46	6.01	2.7	3.31	128.9	129.4	0.5	12.03	17.7	5.67	82.9	86.3	3.4
6	17.2	0	17.2	13.4	6.3	7.1	6.4	-2	8.4	8.29	1.6	6.69	140	153	13	8.7	20.5	11.8	88	88	0
7	16.32	-13.27	29.59	9.8	0	9.8	-1.3	-14.18	12.88	4.5	2.4	2.1	91.5	143.3	51.8	9.94	20.07	10.13	61.6	83.1	21.5
8	7.5	0	7.5	10.1	-1	11.1	4.7	0	4.7	4.7	1	3.7	119.8	134	14.2	10.1	12.3	2.2	76	80	4
9	5	0	5	3	0	3	-2	-10	8	6	1	5	142	159	17	12	17.2	5.2	93	98.8	5.8
10	14.78	9.94	4.84	0	-7.3	7.3	0	-8.15	8.15	6.8	2.2	4.6	80	121.7	41.7	6.7	13.76	7.06	43.4	69.3	25.9
11	16	-6.43	22.43	7.5	-8.18	15.68	6	-8.18	14.18	10.49	5.13	5.36	127.4	148.7	21.3	10.34	20.53	10.19	76.7	86.6	9.9
12	11.8	2	9.8	8.8	-2	10.8	3.4	-2	5.4	3.8	2.86	0.94	122.5	157.6	35.1	17.9	25.6	7.7	92	82	-10
13	7.5	-1.5	9	20.39	3.3	17.09	8.4	7.1	1.3	5.73	3.75	1.98	118.5	142.3	23.8	13.5	23.2	9.7	81.3	100	18.7
14	11.31	-4.2	15.51	14.6	2.87	11.73	7.78	-5.6	13.38	6.75	2.4	4.35	126.9	150	23.1	7.36	15.28	7.92	74.6	76.3	1.7
15	14.6	5.7	8.9	17.6	6.8	10.8	11.27	0	11.27	2.6	2	0.6	94.5	109.9	15.4	16.7	15.67	-1.03	81.6	80.1	-1.5
16	27	8	19	16.8	2	14.8	2.33	-3	5.33	3	6	-3	105.9	131.6	25.7	9	10.9	1.9	80.7	85.2	4.5
17	8.3	-8.1	16.4	5.3	-3.2	8.5	4	-4.8	8.8	6.4	1	5.4	108	143	35	6	16	10	66.6	75	8.4
18	15	0	15	7.1	-2.7	9.8	3.8	-5.4	9.2	5	1	4	138.1	143	4.9	13	22	9	94.7	86	-8.7
19	17.82	8.83	8.99	12	3.6	8.4	11.2	6.4	4.8	10	9.7	0.3	92.5	119	26.5	4.54	12.75	8.21	56	77.2	21.2
20	17.8	0	17.8	16.97	6.92	10.05	17.7	11.24	6.46	6.24	0	6.24	122	129.1	7.1	9.63	26.2	16.57	89.6	96.3	6.7

Preoperative and postoperative CT scan data measured by Observer 3

S.no	wlpre3	wlpos3	change	clpre3	clpos3	change	mlpre3	mlpos3	change	aadpre	aadpos	change	ccapre3	ccapos	change	sacpre3	sacpos	change	ompre3	ompos	change
1	9.5	-10.9	20.4	5.8	-7.5	13.3	0	-13.1	13.1	8.2	6	2.2	116	154	38	9.7	16.2	6.5	61.4	88.4	27
2	12.5	7.8	4.7	6.5	0	6.5	0	-5.2	5.2	9.2	11.2	-2	121	116.4	-4.6	13.5	15.3	1.8	64.8	59.2	-5.6
3	11.1	3.3	7.8	9.1	3.7	5.4	9.4	-4.4	13.8	7.3	1.5	5.8	116	140.5	24.5	7.7	15.5	7.8	80.6	86.9	6.3
4	18.1	5.6	12.5	5.4	0	5.4	2.9	-2.5	5.4	11.7	5.6	6.1	122.6	153.6	31	8.3	14.2	5.9	67.8	95.9	28.1
5	6.8	-4.8	11.6	12.2	-11.5	23.7	4.4	-2.9	7.3	6.4	2.2	4.2	119.3	128.8	9.5	13.2	19	5.8	78.5	88.3	9.8
6	14	0	14	13.1	4	9.1	5.9	-3.8	9.7	8	1.8	6.2	135	146	11	9.5	18.3	8.8	116	91	-25
7	8.5	-13.6	22.1	3.5	-3.5	7	-2	-16.8	14.8	10.3	2.3	8	110	144	34	8.4	21	12.6	66.7	93.6	26.9
8	7.1	-3.7	10.8	8.3	0	8.3	2.4	-7.1	9.5	6.2	1.5	4.7	122	136	14	11	15.3	4.3	75.7	89	13.3
9	6.8	-7	13.8	2.3	-2.5	4.8	-3	-9.5	6.5	5.5	1.7	3.8	159	174.5	15.5	11.9	16	4.1	95.8	106.5	10.7
10	15.2	-12.2	27.4	-0.2	-7.4	7.2	-1.3	-10.6	9.3	4	4.5	-0.5	86.2	124.5	38.3	5.8	10.7	4.9	36	57.4	21.4
11	8.4	0	8.4	4.2	-8.3	12.5	0	-6.8	6.8	11.3	5.9	5.4	131	152.8	21.8	12.5	14.2	1.7	89.9	99.1	9.2
12	8.8	2	6.8	5.1	-3.6	8.7	-1.3	-6.4	5.1	4.4	2	2.4	127.6	149.5	21.9	16.3	22	5.7	64.2	87.2	23
13	3.9	0	3.9	3.1	0	3.1	-3.9	-7.7	3.8	6.5	5.2	1.3	124.8	144.5	19.7	15.7	19	3.3	86.9	85.7	-1.2
14	9.1	-2.7	11.8	9.3	3.2	6.1	3.2	-5.2	8.4	6.4	2.5	3.9	142.7	161.6	18.9	7.9	12.1	4.2	85.8	73	-12.8
15	19.2	5.2	14	19.5	3.4	16.1	14.2	-1.6	15.8	3.9	6.2	-2.3	115.1	113.4	-1.7	9.3	13	3.7	76.8	80.7	3.9
16	26.7	6.7	20	16.2	2.3	13.9	11.6	-2.3	13.9	3.6	6	-2.4	112.5	135.2	22.7	9.8	14.7	4.9	84.8	91.6	6.8
17	5.6	-7	12.6	6.6	-2.1	8.7	2.9	-3.3	6.2	6.8	1	5.8	116.7	150.7	34	7.61	18	10.39	65	89.4	24.4
18	11.2	0	11.2	2.7	-4.8	7.5	0	-7.6	7.6	6.4	2.2	4.2	146.1	151.5	5.4	15.4	23.5	8.1	100.7	91.4	-9.3
19	19.4	8.6	10.8	5	0	5	4.7	-2.8	7.5	13	11.3	1.7	99.5	130.3	30.8	5.4	14.3	8.9	51.5	75.5	24
20	14.8	4.6	10.2	15.3	7.3	8	13	0	13	5.8	1	4.8	129.2	153.7	24.5	9	24.7	15.7	99.3	99.1	-0.2

Appendix 3

Preoperative and follow-up craniometry of all the patients by Observer 1.

S.no	wl pre	wl f/up	W L 1	cl pre 1	cl f/up	C L 1	ml pre	ml f/up	M L 1	aad pre	aad f/u	AAD 1	sac pre	sac f/up	SAC 1	cca pre	cca f/up	CCA 1	ome pre	ome f/u	OME 1
1	4.65	-6	10.65	4.65	-2	6.65	-0.7	-3	2.3	5.9	5	0.9	8.8	17	8.2	109.1	135	25.9	56.8	84	27.2
2	15.1	0	15.1	5.9	0	5.9	0	-3	3	13	4	9	9	16	7	103.3	140	36.7	70.6	84	13.4
3	12.4	6.8	5.6	16.2	7	9.2	9.1	0	9.1	9.6	3.8	5.8	10.2	17	6.8	133.3	142	8.7	96.5	90	-6.5
4	6.9	0	6.9	12.2	0	12.2	2	0	2	6.9	1	5.9	11.5	13.8	2.3	107.8	125.2	17.4	77.9	75.5	-2.4
5	13.2	16	-2.8	0	-3.8	3.8	0	-2	2	4.04	8	-3.96	6.8	8.4	1.6	72.8	97	24.2	32.7	54	21.3
6	16.6	0	16.6	5.6	-4	9.6	1.7	-3	4.7	12.46	5.8	6.66	13.2	17	3.8	129.4	131.6	2.2	81.6	90.4	8.8
7	9.3	4	5.3	9.4	0	9.4	3.7	-4	7.7	5.4	2	3.4	17	21	4	123.3	141.4	18.1	65.3	80	14.7
8	4	0	4	13.7	0	13.7	4.5	-6	10.5	6.5	5	1.5	14	16.8	2.8	124.8	142	17.2	85	90	5
9	10.7	0	10.7	12.7	-2	14.7	5	-3	8	6.3	2	4.3	8	13	5	132.6	155.4	22.8	69.9	87	17.1
10	6.6	-1.2	7.8	6.6	0	6.6	4.6	4.7	-0.1	8.8	0	8.8	9	21.9	12.9	79.4	149	69.6	65.7	74	8.3
11	9.5	0	9.5	6	0	6	0	-6	6	7.7	3	4.7	15.9	20	4.1	137.6	142.8	5.2	90.7	90	-0.7
12	15.4	0	15.4	11.14	8	3.14	8.2	5	3.2	4.9	0	4.9	8.7	20	11.3	133	135	2	92.4	96.7	4.3
13	26.5	14.7	11.8	17.5	1.2	16.3	13.9	0	13.9	5	3.9	1.1	10.3	12.5	2.2	105.3	124.1	18.8	79.3	82.5	3.2
14	17.7	9	8.7	10.6	12.7	-2.1	8.2	4	4.2	17.4	13.9	3.5	8.1	13.9	5.8	88.9	110	21.1	52.5	54.1	1.6
15	7.96	0	7.96	4.95	-1.7	6.65	5.6	-9.9	15.5	4.5	0	4.5	12.4	18	5.6	150.3	180	29.7	98.7	109.8	11.1
16	11.36	3.7	7.66	8.45	0	8.45	9.9	3.6	6.3	5.24	2.9	2.34	9.2	18	8.8	119.3	145	25.7	78.6	79	0.4
17	12.36	2	10.36	7.88	2.7	5.18	5.58	-6.4	11.98	12.6	7	5.6	12.44	9.8	-2.64	100.5	109.2	8.7	63.9	62	-1.9

Preoperative and follow-up craniometry of all the patients by Observer 2.

S.no	wl pre	wl f/up	W L 2	cl pre	cl f/up	C L 2	ml pre	ml f/up	M L 2	aad pr	aad f/	AAD 2	sac pre	sac f/u	SAC 2	cca pre	cca f/u	CCA 2	ome p	ome f/	OME 2
1	6.04	-2	8.04	6.5	-2	8.5	0	-4	4	4.3	4	0.3	7.14	18.4	11.3	95.3	142	46.7	62.8	80.6	17.8
2	15	1	14	9.37	1.2	8.17	3.7	0	3.7	11.3	5	6.25	5.6	15.8	10.2	112	144	32.5	71.2	85.6	14.4
3	17.2	8.1	9.1	13.4	10	3.4	6.4	2	4.4	8.29	4.3	3.99	8.7	17	8.3	140	144	4	88	85.1	-2.9
4	7.5	1	6.5	10.1	3	7.1	4.7	0	4.7	4.7	1	3.7	10.1	12.8	2.7	120	129	8.9	76	76.2	0.2
5	14.8	14.6	0.21	0	-4.3	4.3	0	0	0	6.8	8.14	-1.34	6.7	6.9	0.2	80	94.9	14.9	43.4	55.5	12.1
6	16	0	16	7.5	-2	9.5	6	-2.8	8.8	10.5	6.44	4.05	10.3	20	9.63	127	132	4.5	76.7	85.4	8.7
7	11.8	4.8	7	8.8	0	8.8	3.4	0	3.4	3.8	2.8	1	17.9	27	9.1	123	143	20.6	92	75.7	-16.3
8	7.5	-1	8.5	14.8	0	14.8	8.4	-5	13.4	5.73	4	1.73	13.5	17.5	4	119	140	21.5	81.3	92.6	11.3
9	11.3	-4.1	15.4	14.6	-4.5	19.1	7.78	-4.7	12.5	6.75	2.1	4.65	7.36	15.4	8.04	127	143	16.1	74.6	76.9	2.3
10	8.3	0	8.3	5.3	0	5.3	4	0	4	6.4	3.37	3.03	6	14.5	8.5	108	130	21.5	66.6	75.4	8.8
11	15	0	15	7.1	0	7.1	3.8	-4.5	8.3	5	2	3	13	22	9	138	136	-2.1	94.7	90.1	-4.6
12	17.8	0	17.8	17	7.2	9.77	8.7	9	-0.3	6.24	0	6.24	9.63	22.9	13.3	122	130	8.2	89.6	95.2	5.6
13	27	13.6	13.4	16.8	5	11.8	12.33	0	12.3	3	4.3	-1.3	9	11	2	106	117	11.1	80.7	82.8	2.1
14	12.5	9.5	3.04	8.82	7.7	1.12	0.98	0	0.98	7.84	12	-4.11	13.8	13	-0.82	106	101	-4.6	63.6	60.9	-2.7
15	5	2	3	3	0	3	-2	-9	7	6	0	6	12	17.5	5.5	142	164	22	93	101	8
16	10.4	4	6.38	11.1	0	11.1	11.53	3	8.53	6.1	3	3.1	7.69	18	10.3	106	143	36.9	77.3	80	2.7
17	16.3	2	14.3	9.8	3	6.8	-1.3	-6	4.7	4.5	6.8	-2.3	9.94	9	-0.94	91.5	110	18.5	61.6	70	8.4

Preoperative and follow-up craniometry of all the patients by Observer 3.

S.no	wl pre	wl f/u	W.L 3	cl pre	cl f/up	C L 3	ml pre	ml f/u	M L 3	aad pr	aad f/	AAD	sac pr	sac f/u	SAC 3	cca pr	cca f/u	CCA	ome p	ome f/	OME
1	9.5	0	9.5	5.8	-3.6	9.4	0	-3.6	3.6	8.2	6.3	1.9	9.7	18.2	8.5	116	141	25.2	61.4	90	28.6
2	18.1	0	18.1	5.4	0	5.4	2.9	-4.3	7.2	11.7	5.3	6.4	8.3	16.8	8.5	123	143	20.3	67.8	85.2	17.4
3	14	0.3	13.7	13.1	5.2	7.9	5.9	0	5.9	8	3	5	9.5	15.3	5.8	135	143	8	116	88.3	-27.7
4	7.1	-2.9	10	8.3	2.4	5.9	2.4	-4.4	6.8	6.2	3.6	2.6	11	13.8	2.8	122	129	6.8	75.7	82.7	7
5	15.2	16.3	-1.1	-0.2	-5.2	5	-1.3	-9.1	7.8	4	8.1	-4.1	5.8	8.1	2.3	86.2	102	15.8	36	48.9	12.9
6	8.4	6.9	1.5	4.2	-5.6	9.8	0	-5.1	5.1	11.3	9.8	1.5	12.5	14.8	2.3	131	134	2.6	89.9	90.1	0.2
7	8.8	4.8	4	5.1	0	5.1	-1.3	-6.9	5.6	4.4	2.4	2	16.3	18.5	2.2	128	145	17.4	64.2	76.6	12.4
8	3.9	0	3.9	13.1	0	13.1	3.9	-9.2	13.1	6.5	5	1.5	15.7	16.6	0.9	125	139	14.2	86.9	83.3	-3.6
9	9.1	-3.4	12.5	9.3	0	9.3	3.2	-6.3	9.5	6.4	3	3.4	7.9	12	4.1	143	160	17.3	85.8	82.8	-3
10	5.6	-3.4	9	6.6	0	6.6	2.9	-3	5.9	6.8	3.2	3.6	7.6	14.6	7	117	138	21.3	65	68.8	3.8
11	11.2	1.9	9.3	2.7	0	2.7	0	-5.7	5.7	6.4	4.6	1.8	15.4	22.9	7.5	146	144	-2.6	101	95.5	-5.2
12	14.8	0	14.8	15.3	8.6	6.7	7	3.6	3.4	5.8	2.2	3.6	9	22.6	13.6	129	145	15.7	99.3	97.5	-1.8
13	26.7	13.3	13.4	16.2	2.2	14	11.6	0	11.6	3.6	3.4	0.2	9.8	10.9	1.1	113	124	11.6	84.8	80.3	-4.5
14	12.5	9.2	3.3	6.5	14.2	-7.7	0	5.3	-5.3	9.2	13.9	-4.7	13.5	13.5	0	121	105	-15.6	64.8	51.1	-13.7
15	6.8	0	6.8	2.3	-1.3	3.6	-3	-9.9	6.9	5.5	0	5.5	11.9	18.8	6.9	159	172	13	95.8	10.4	-85.4
16	11.1	4.4	6.7	9.1	0	9.1	9.4	3.4	6	7.3	3.2	4.1	7.7	17	9.3	116	140	24	80.6	82	1.4
17	8.5	1.9	6.6	3.5	3.9	-0.4	-2	-5.3	3.3	10.3	6.8	3.5	8.4	9.2	0.8	110	106	-4	66.7	61.2	-5.5

CRANIOVERTEBRAL REALIGNMENT FOR BASILAR INVAGINATION

Objective:

To assess the clinical outcome, radiological realignment following surgery for basilar invagination.

Methods and materials:

Seventeen basilar invagination and three basilar impression patients included from 2007-2009. Neurological status assessment, X-ray cvj and CT cvj was done preop, after 6 months and yearly postoperatively. Craniometry was be subjected to statistics and significance assessed based on Wilcoxon signed-rank test. Craniometry findings were evaluated by 2 neurosurgeons and 1 neuroradiologist and interclass correlation coefficient was obtained.

Results:

Cvj realignment maintained at 19/20 patients. 17 out of 20 patients had a mean follow-up of 13.1±5.2 months. The mean Nuricks grade improved from 3.2±1.2 to 2±1.2 (p value=0.002) postoperatively and modified JOA score improved from 11.1±3 to 14.7±2.2 (p value=.000). The mean levels of the odontoid tip above Wackenheim's Clival line on preoperative, postoperative and at follow-up scans are 11.9± 5.2, -0.81± 5.0and 2.97± 5.6 respectively. The mean level of odontoid tip in relation to Chamberlain's line at on preoperative, postoperative and at follow-up scans are 8.60± 4.3, -0.43± 4.0and 1.23± 4.28 mm respectively and a mean change of 9 ± 2.7 mm was achieved due to surgery. It was seen that the odontoid was 3.9 ± 5.7 mm above Mac Rae's line preoperatively, while in the immediate postoperative period it was 4.7 mm below this line. It was shown that the mean value of atlantoaxial distance before surgery was 7.1 ± 2.4 and

it was 3.2 ± 2.7 mm postoperatively and it is increased to 4.3 ± 3.2 mm at follow-up .The mean value of sagittal canal diameter before surgery was 10.5 ± 2.9 and it was increased to 18.5 ± 4 mm postoperatively and it is decreased to 16 ± 4 mm at follow-up. The mean value of clival canal angle before surgery was 119 ± 17 degrees and it was increased to 143.1 ± 12.8 degrees postoperatively and it is decreased to 134 ± 18 degrees at the follow-up. The mean value of Modified Omega angle before surgery was 76.3 ± 16 degrees and it was increased to 86 ± 11.8 degrees postoperatively and it is decreased to 79 ± 12.1 degrees at follow-up. All these value changes at the follow-up were statistically significant except modified omega angle. ICC showed good correlation between 3 observers except the measurement of modified omega angle at the follow-up scan. One of our patients had a vertebral artery injury during the surgery and one patient had a resurgery due to implant related complication.

Conclusion:

Craniovertebral realignment provides excellent neural decompression and clinical outcome in patients with basilar invagination. It is a technically demanding surgery and accurate placement of the implants plays a vital role in the reduction of basilar invagination. Although good bony fusion is seen by 6 months, short term follow-up indicates some settling of graft and spacer. Therefore longer follow-up is mandatory before considering C1 2 distraction surgery as a gold standard treatment for basilar invagination.